Orthopedic Surgery
in
Infancy and Childhood

Contributors

JOHN SPEER DONALDSON, B S , M D

Assistant Professor Orthopedic Surgery University of Pittsburgh.
Senior Staff Children s Presbyterian and Shadyside Hospitals, Pittsburgh.
Consultant in Orthopedic Surgery Industrial Home for Crippled Children, Magee Hospital, Pittsburgh and U S Veterans Hospital Oakland, Pa

WILLIAM F DONALDSON, M.D , F.A.C.S

Assistant Instructor Orthopedic Surgery University of Pittsburgh
Associate Staff Children s and Presbyterian Hospitals.
Attending Physician, U S Veterans Hospital Oakland, Pa.

ALBERT BARNETT FERGUSON, SR , M.D

Associate Professor of Orthopedic and Fracture Surgery Boston University
Consultant in Orthopedic Roentgenology, Children s Medical Center Boston, Memorial Hospitals, Lakeville Sanitarium, Massachusetts Crippled Children s School.
Roentgenologist, Brockton Hospital, Brockton, Massachusetts.

ROBERT KLEIN, M.D

Associate Professor of Pediatrics University of Pittsburgh
Markle Scholar in Medical Sciences.
Associate Staff Children s Hospital, Pittsburgh.

FRANK H STELLING, M D , F.A.C.S

Chief Surgeon Shriner s Hospital for Crippled Children, Greenville S C
Instructor Orthopedics, Duke University School of Medicine, Durham, N C
Chief Orthopedic Surgery Greenville General Hospital Greenville S C
Orthopedic Surgeon S C State Crippled Children s Services
Consultant Orthopedic Surgeon Donaldson Air Force Base, Greenville, S C

Orthopedic Surgery in Infancy and Childhood

By Albert Barnett Ferguson, Jr, B A, M D

Silver Associate Professor Orthopedic Surgery University of Pittsburgh
Chief of Orthopedic Service University of Pittsburgh Medical Center
Senior Staff
Children's, Presbyterian and St Margaret's Hospitals Pittsburgh
Chairman Medical Advisory Board
Harmarville Rehabilitation Center
Consultant, Orthopedic Surgery
U S Veterans Hospital Oakland and Green County Memorial Hospital, Waynesburg



THE WILLIAMS & WILKINS CO
BALTIMORE 1957

Preface

This volume is the outgrowth of that period in life when the subject matter of orthopedic surgery was laboriously amassed by running from text to text and article to article Excellent though so many volumes are, none of them appeared to have gathered under one cover details regarding the many odd syndromes encountered in the practice of orthopedic surgery dealing with infants and children. Pediatricians and residents have often indicated by their questions that they were undergoing a similar experience

No syndrome, photograph or roentgenogram is presented which was not personally seen by the author No feature of disease is reiterated which has not been a feature in our own experience

Obviously no text arises without roots in the past, and the excellent volumes of Colonna Howorth, Mercer, Platt Wiles, Fairbank, Shands, Campbell, Luck Smillie and DePalma have also served as references This text does not include a consideration of fractures, for the subject is adequately covered in Walter Blount s recent book, *Fractures in Children*

If clarity in visualizing these syndromes has been achieved it is a reflection of the teaching of Joseph Barr, William T Green and Albert B Ferguson, Sr They are not responsible for what others may deem as errors, however

Certain areas in the book have been done by others Frank Stelling has contributed his knowledge of reconstruction of the hand to the section on the upper extremity Scoliosis is the work of William Donaldson of Pittsburgh. John Donaldson of Pittsburgh has added the chapter on the neck. Robert Klein encompasses rickets and scurvy from the metabolic point of view Albert Ferguson, Sr writes of defective formation of bone from cartilage All these authors are deserving of my humble thanks in enabling a sound production of a book on orthopedic surgery in the infant and childhood years

The Children s Hospital of Pittsburgh is blessed with men of selfless spirit whose help has been invaluable This is particularly true of Bertram Girdany, George Fetterman and Thomas Brower although no one failed to respond when called upon The pleasant and productive atmosphere of this hospital is a reflection of its guiding spirit and medical director Edmund McCluskey

The manuscript has been typed, retyped and retyped again by my secretary already overloaded with budgets, bills and records This type of dedication is typical of Mary Cosgrove.

My children and my wife, Louise, have taken night work which deprived them of companionship, with good spirit. My wife s patient understanding is not

found often in this world—without it such an arduous task could not be completed.

The drawings are the result of the talents of Margaret Croup except for those accompanying the section on the upper extremity. The photographs and roentgenogram reproductions were done by Albert Levine and James Stark. The patient cooperation exhibited by these worthy individuals has been a source of pleasure and has eased the task immeasurably.

The publishers and particularly Dick Hoover have been patient and exhibited the skills of their profession with consummate ease.

Great care and effort have been used to duly credit thoughts, drawings and photographs whose origin might in any way be elsewhere. Should any omission have arisen, it is entirely inadvertent and not by conscious design.

The regional arrangement of the text will, we hope, make reference easy and invite a return visit.

A. B. F., Jr.

Contents

1

The Foot

FLAT FEET IN CHILDHOOD

Perhaps the most common orthopedic problem confronting the physician caring for the growing child is flat feet. The symptoms that may be associated with weak feet are often not recognized The child who appears lazy, dislikes long walks, and comes indoors while the rest of his age group is still out playing may be suffering from foot strain The toeing in or "pigeon toeing" in a child's gait is frequently due to weak feet. When actual pain is complained of, it frequently is located not in the foot but in the calf This leg discomfort has two characteristics it is intermittent, it tends to occur at the end of the day

On first standing, the infant s feet are almost invariably pronated, spread apart in a wide-based stance for balance as he grips his support. Standing barefoot in this position will stretch out the ligaments of the medial side of the foot. Gradually he brings his feet closer together, with supination of the feet. The infantile fat is redistributed as the child's activity increases As a preventive measure the child should be in firm shoes with a heel from the onset of his gaining upright posture The child whose foot is not responding to this increased activity is ordinarily not recognized until the age of two or over

Clinical Picture

The child is brought to medical attention because the parents have noted the flat or pronated feet, or the child toes in (*i e*, walks pigeon toed), or the patient limps or complains of foot or leg pain.

A history of intermittent limp with an essentially negative physical examination except for pronated feet suggests foot strain. Calf pain, when due to muscle fatigue from supporting an inadequate foot tends to be located in the region of the posterior tibial muscle. Foot pain due to ligamentous strain is located by the patient along the medial arch.

Physical Examination

The examiner seeks to find, not a flat foot, but a foot which is functioning in a position which throws strain on its ligaments and muscles and does not use its bony support. The degree of arch medially and longitudinally is noted

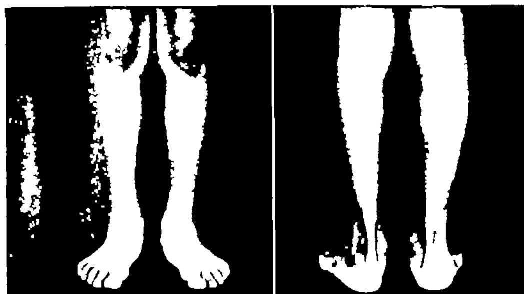


Figure 1 Pronated feet with prominence of the calcaneo-navicular area medially and a valgus position of the os calcis seen in the posterior view

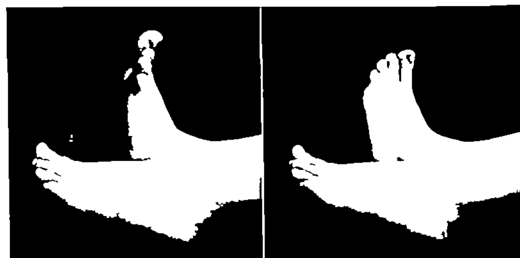


Figure 2 The foot everted (left) will dorsiflex further than the foot inverted (right)

in recumbency Most feet are well arched in this position but some feet are architecturally flat on non weight bearing. The tightness of the heel cord is noted by dorsiflexing the foot in inversion while the knee is straight. A foot normally should be able to dorsiflex at least ten degrees above the right ankle when examined in this way

A tendency to hyperextended knees elbows and thumbs is noted as giving a clue to the ligamentous laxity of the child. The ability of the foot to roll from

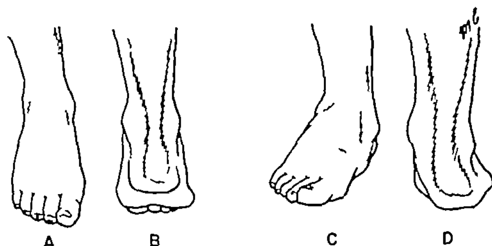


Figure 3 The foot with ligamentous laxity is normally constituted without weight bearing (A and B) With weight bearing the foot pronates and the os calcis goes into a valgus position (C and D)

full eversion to full inversion should be noted. The inability to invert the foot may be due to peroneal spasm which can be confirmed by palpating the peroneal muscles as the inversion is attempted In spasm, they will stand out as taut structures limiting inversion An examination of the complete child, with special attention to all joints, is carried out in every case of suspected foot strain.

When the child is standing the position of the os calcis in relation to the weight bearing line is of importance The degree to which the heel slopes laterally carrying the weight bearing line out from the tibia is the degree of pronation of the foot. This is best noted from the posterior view with the foot at eye level As the os calcis everts beneath the astragulus the medial side of the foot comes into prominence Pressure on the medial side of the foot will reveal whether the os calcis can still be tilted into an upright position The contrast between the weight bearing and non weight-bearing position is important from the standpoint of strain.

The alignment of the leg will affect the foot Tibia vara with a medial facing of the ankle mortise will mean that the foot must pronate in order to bring its weight bearing surface on the ground. The internal torsion of the tibia may be confused with the tibia vara deformity or may be part of it. Internal torsion is noted by placing the knee facing directly to the examiner and dorsiflexing the foot in line with the ankle joint. It should be in the same plane as the knee. Deviation of the ankle joint medially in relation to the plane of the knee joint may be due to internal torsion. A facing outward or laterally of the coronal plane of the ankle joint in relation to the knee is external torsion

Internal torsion accentuates a bow legged appearance, but gives rise to "in toeing " a somewhat stronger weight-bearing position than that produced by external torsion. Here the foot turns outward carrying it away from direct transmission of body weight and tending to accentuate pronation

Valgus at the knee frequently develops in children with pronated feet This is particularly true in youngsters with chubby thighs whose legs are widely

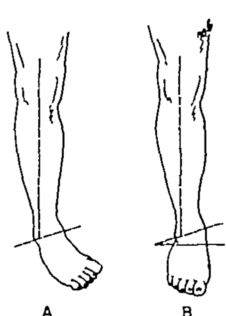


Figure 4 In tibia vara there is a medial facing of the distal tibial surface at the ankle joint, resulting in the necessity for the foot to pronate to bear weight on its medial side.

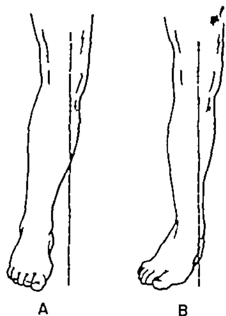


Figure 5 The weight bearing line is brought medially by valgus at the knee as in A External torsion of the tibia rotates the foot outward and away from the weight bearing line B

separated on weight bearing. Valgus may be a product of the same relaxed ligamentous structure that allows the foot to pronate.

The Gait

One of the most important features of childhood foot strain is the gait. 'In toeing or walking "pigeon toed is frequently a protective gait to guard against foot strain. There may of course, be mechanical factors in the leg such that no other form of gait is possible, but on the basis of frequency a protective gait is much more likely Nor is every child who toes outward without difficulty Such a gait will tend to develop a pronated foot in a growing child.

Treatment

The placing of wedges on the lateral side of the shoe to make an in toeing child toe outward is contraindicated. This pronates the foot still further, increases the strain and often results in increased in toeing to compensate.

The treatment of weak mechanical feet consists of providing adequate support to hold the foot in a strong position. Growth taking place while the foot is held in this position should result in relative tightening of the ligaments and the prevention of bony deformity It is not possible to make the worst pair of feet into the strongest but it should be possible to sufficiently change the foot mechanics so that they do not constitute a point of great weakness as the individual attempts to walk and stand in life. The failure of inadequate treatment should not result in wholesale condemnation of the method.

A medial arch pad or support helps to prevent rotation of the foot within

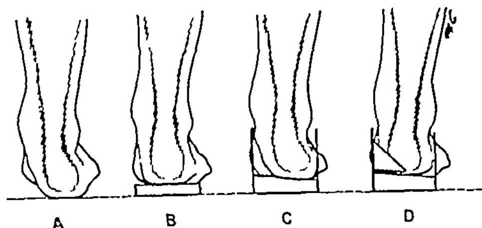


Figure 6 The effect of various measures on the position of the os calcis is shown In B a normal heel alone improves the position but this effect is increased further by medial wedge combined with a longitudinal arch pad A medial wedge alone is not sufficient to keep the heel from the valgus (C) but combined with a pad the full effect of both is felt. (D)



Figure 7 The effect of a heel alone on foot position. Valgus of the os calcis is largely corrected.

the shoe This increases the effectiveness of a medial heel wedge which is designed to tilt the hindfoot into inversion. The usual well tolerated medial heel wedge height is one-eighth inch. If the hindfoot is controlled, the forefoot will follow Medial arch supports that are carried too far foreward will result in difficulty in fitting the foot over them into the shoe.

The sub astragular area is the point where support is needed most The usual height for a medial arch support in childhood is three-eighths of an inch. A medial prolongation of the heel may be added in addition to the wedge This so-called 'Thomas" heel is a part of shoes that are sold as ' orthopedic shoes.

Straight-last shoes (Plumb-line) disguise in toeing by the child and are used where the forefoot tends to turn medialward or adduct in relation to the hind foot.

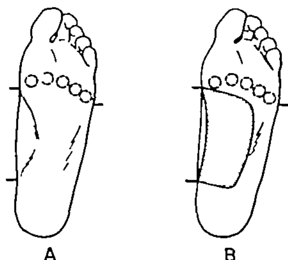


Figure 8 In tracing a foot for medial arch support the area just proximal to the first and fifth metatarsal head is marked as well as the posterior termination of the medial arch. Keeping the anterior end of the pad behind these prominences is important for comfort and for fitting the pad into the shoe The pad is brought well toward the middle of the foot posteriorly to support the subastragular area.



Figure 9 Leather arch supports with felt insert. The extra felt is inserted when the first becomes compressed.

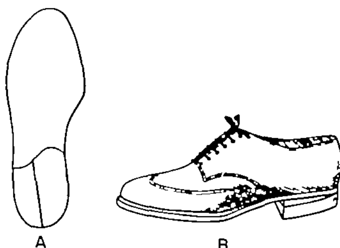


Figure 10 Inner heel wedge location to aid in inverting the os calcis.



Figure 11 In heel cord stretching in the older child the heel of the posterior limb is kept on the ground The elbows are flexed and the child leans toward the wall

Exercises for the feet of an inverting or supination type have been of little help in the author's treatment of flat or pronated feet in childhood. Adults derive much more benefit from them.

There is one exception to this the case of a tight heel cord. Since the foot can dorsiflex further in inversion the youngster is forced into a weak mechanical position in order to get his heel on the ground in the presence of tight heel cords No progress in the direction of improvement of the feet can be expected while the heel cord remains tight The mother can stretch this tendon when the child is small by inverting and dorsiflexing the foot with the knee extended When the child is older he or she can be taught heel cord stretching exercises as illustrated. Such exercises need not be done to the point of boredom, but should be regularly and thoroughly done at least once each day It usually takes three to four months to exhibit improvement.

Correction of a foot and gait problem will not take place instantaneously when corrective measures are applied, but only with further development and growth of the foot.

Stiff plates of steel or plastic tend to make the foot rigid and wed the child permanently to their use Flexible supports appear preferable Pasted in longitudinal arch supports such as are commonly seen commercially as a general rule are inadequate since a fully adequate support will necessarily give the child some slight difficulty in getting his foot into the shoe.

This problem is avoided by not building the pad too far forward Its most important function in the simple pronated foot of childhood is to control the hindfoot the forefoot must necessarily fall in line A longitudinal arch pad

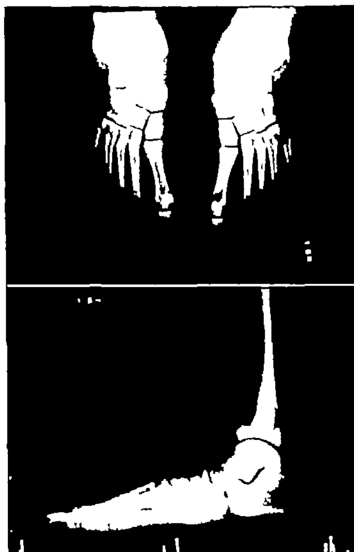


Figure 12 Roentgenograms of disabling flat feet See also Figure 13

ends behind the first metatarsal head and extends posteriorly to the end of the arch and inward (laterally) from there to tilt the os calcis into an upright position by its support

DISABLING FLAT FEET

HYPERMOBILE FLAT FOOT WITH SHORT TENDO-ACHILLES

Harris and Beath have reemphasized a type of flat foot frequently associated with disability The disability is often not apparent until the "teen" age is reached This foot has excessive ligamentous laxity and a short tendo-achilles which are recognizable on physical examination. The foot on non weight bearing appears to have a good longitudinal arch both medially and laterally On standing the foot pronates severely

This foot combines two elements likely to cause disability (1) Function

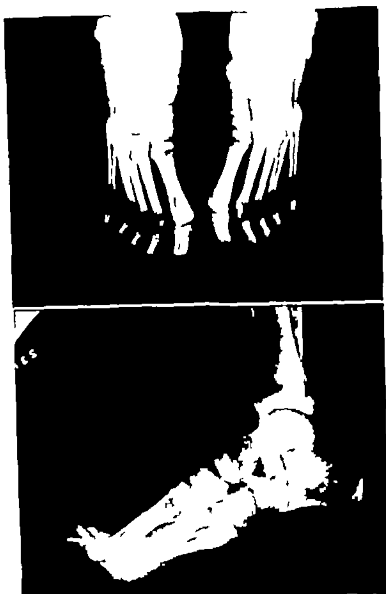


Figure 13 The foot as in Figure 12 treated with subastragalar bone block The os calcis has been brought under the astragalus in both AP and lateral views

in a position for which the foot was not designed (2) A mechanical abnormality which forces use in this position, although not necessarily a primary cause. Harris felt that the primary cause was a structural abnormality, the head of the talus being displaced downward and inward in relation to the os calcis It was noted in Canadian soldiers once in every 145 men examined

Since the severity of this condition varies greatly, treatment will vary from mere support and exercises to operation Operation is beyond the scope of this book since is would be used only in severe cases demonstrating incapacitating disability in the adult. Stretching of the tendo-achilles is important, and in the "teen" age group metal type supports perhaps including a lateral flange as in the Whitman plate may be used. It is hard to get the foot well supported here and a plaster mold of the foot is usually made with the foot in a corrected position as a guide to the making of the support.



Figure 14 The hypermobile flat foot with short tendo achilles results in severe pronation

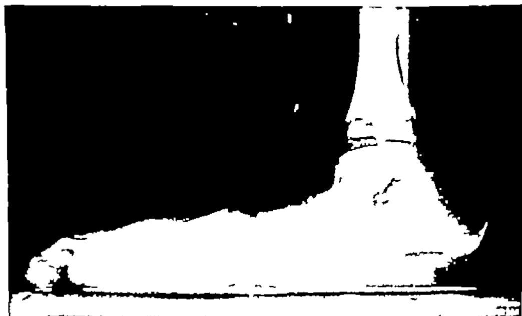


Figure 15 Lateral roentgenogram of the foot weight bearing showing the plantar flexed talu depressed na icular loss of the medial and lateral arches of the foot and subluxation of the talar l d over the anterior portion of the os calcis.

THE PLANTAR FLEXED TALUS

A type of foot seen occasionally in childhood is one in which the astragalus is in a fixed equinus position Of necessity, weight bearing in this foot must result in eversion and abduction of the forefoot and the rolling out of the os calcis as the entire foot rotates around the fixed talus

The foot can be recognized on clinical examination by bringing the foot back underneath the talus and finding the foot then unable to dorsiflex. The head of the astragalus is often actually weight bearing with a callus over the area of the prominent astragalar head.

Such a foot is brought into good alignment and then wedged upward by a wedging cast to get the foot into adequate dorsiflexion There is a pronounced tendency to recurrence rarely, when conservative measures fail, operation may be justified The subastragalar bone block described by Grice for paralytic flat feet has been found satisfactory by the author A triple arthrodesis such as described by Hoke results in a stable foot, but is a more extensive procedure

PERONEAL SPASTIC FLAT FOOT

Peroneal spastic flat foot is recognized clinically by an inability to invert the foot. On attempted inversion the peroneal muscles will be noted standing out as taut structures Lapidus notes that rarely the extensor digitorum may join the peroneal in holding the foot in pronation

There are a number of possible causes for this condition. In children inflammatory lesions such as tuberculosis may be present in the mid or subtarsal area. Subtalar arthritis of a rheumatoid type may be present, and lesions such as osteoid osteoma have been found Occasionally the condition may be seen secondary to trauma It is important to rule out bony anomalies such as calcaneo-navicular coalition which may give rise to midtarsal strain and form the most common etiologic group

The foot may be rigidly fixed regardless of any contracture of the peroneals In those states where there is a congenital fusion of the tarsal bones, motion into inversion is obviously impossible regardless of the guarding action of the peroneals.

Harris and Beath found the most significant and frequent anomaly to be a talo-calcaneal bridge rather than a calcaneo-navicular bar, and are responsible for its clear recognition. Such a bridge is apparently a variation similar to that which produces the os sustentaculi, and may actually represent fusion of this accessory bone at either end A similar bone, calcaneus secondarius, is the basis of the calcaneo-navicular bar

The bridge may be bony or fibrous The limitation of subtalar joint motion causes a characteristic talo-navicular infringement. This gives rise to a characteristic dorsal lipping of the talus. A forty five-degree postero-anterior view of the os calcis is necessary to demostrate the subtalar joint and its pathology

Treatment

Peroneal spastic flat feet must be thoroughly investigated as to etiology The condition usually persists until the underlying cause is cured. This sometimes

requires a triple arthrodesis. Rest in bivalved casts with daily exercise (non weight bearing) is part of the treatment of arthritic conditions. This is followed by support of the foot in normal weight-bearing position. Dickson, Dively and Whitman recommend manipulation under anesthesia and immobilization in plaster followed by physiotherapy

The underlying etiologic factor usually must be cured to prevent a recurrence of the protective muscle spasm. Local excision of accessory bones and bars in the cartilage state has been attempted in childhood with some success.

METATARSUS ATAVICUS

In the metatarsus atavicus anomaly described by Morton the first metatarsal is excessively short. The length is usually less than that of the second and third. This decreased length results in a tendency to pronate the foot in order to get the head of the first metatarsal into a weight bearing position. Feet pronated for this reason should be supported by a prolongation of a longitudinal arch pad beneath the first metatarsal to procure weight bearing.

METATARSUS PRIMUS VARUS

Metatarsus primus varus is seen at birth. The first metatarsal is adducted deviating medialward so that a well-defined defect exists between the first and second metatarsal head

This results in abnormal weight bearing on the second metatarsal and the use of footwear will carry the first toe into a hallux valgus position. The hallux valgus deformity is the most striking physical aspect.



Figure 1[illegible] A metatarsus primus varus. The medial deviation of the first metatarsal is seen with the great toe lying in a valgus position.

When the deformity is severe enough to cause abnormal weight bearing and deformity of the toe, an osteotomy of the base of the first metatarsal may be done to bring it into normal alignment with the other metatarsals. Such an osteotomy, to be successful, should be oblique with the long edge of the proximal portion on the medial side.

PAINFUL HEELS IN CHILDREN

Pain located at the superior angle of the os calcis is a common childhood complaint in the 6- to 12-year old age group. The cause of such complaints is varied.

The principal cause appears to be excessive prominence of the posterior superior angle of the os calcis. Such a prominence results in the formation of a callus in this area as a result of pressure from the heel of the shoe. Relief from this discomfort is obtained by removing the stiff heel counter from the shoe and obtaining a make in which the upper of the shoe has sufficient height so that the gripping pressure of the heel is above the prominence.

Tendinitis of the achilles tendon is revealed by tenderness over this tendon and occasionally swelling or crepitus noted on palpation. The roentgenogram reveals thickening of the achilles tendon shadow. Complete rest, which is readily achieved by a plaster cast immobilization, may be indicated until symptoms disappear. Elevation of the heel and hydrocortone injections may relieve the milder cases.



Figure 1 The normal development of the os calcis includes a dense stage of the apophysis. Left 4½ years. Middle 5 years. Right 8 years.

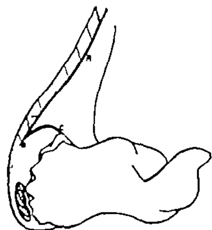


Figure 18 Diagram emphasizing the angle made by the junction of the tendon achilles and os calcis. This angle is obscured by inflammatory swelling at the heel. The angle AOC is formed by the junction of the achilles tendon and superior surface of the os calcis.

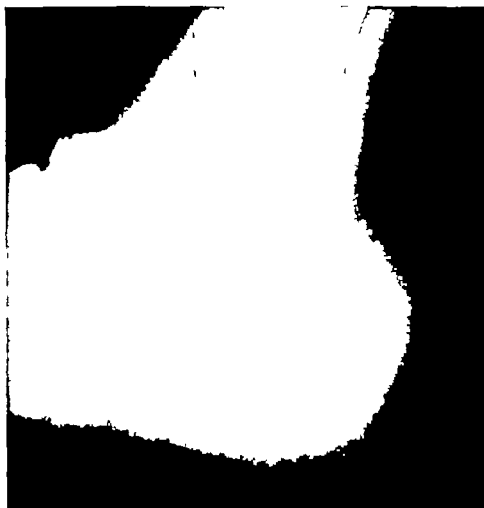


Figure 19 Inflammatory swelling about the heel in osteomyelitis of the os calcis. This marked inflammation obscures the angle AOC.

Bursitis of the achilles bursa beneath the achilles tendon rarely occurs. Such a lesion may form part of the calcaneal apophysitis syndrome, which has been felt in the past to be a process of osteochondritis similar to that of Kohler's disease Freiberg's disease and Legge-Perthes' disease. Such a diagnosis was based on the density of the apophysis A review of the development of the os calcis from infancy reveals however that the calcaneal apophysis normally passes through an extremely dense stage which is alleviated after union with the parent bone. No unusual fragmentation has been noted.

Roentgenograms have revealed changes in those cases which clinically appeared to have calcaneal apophysitis with tenderness and thickening at the junction of the achilles tendon with the bone. These changes have consisted of thickening of the soft tissue shadow in this area and in the angle formed by the juncture of the tendon and the os calcis. Rarely there is ossification of the apophysis at some distance from the parent center



Figure 20 Aseptic inflammatory swelling at the junction of achilles tendon and os calcis

Both these changes and the syndrome itself has been quite rare in our experience.

Treatment

Elevation of the heel and softening of the heel counter and a changing of its height have all helped to alleviate symptoms. Rest in a plaster cast, hydrocortone injection and heel cord stretching may be of some value in the more resistant cases.

METATARSUS VARUS

In pediatric orthopedic practice two variations of the common deformity, metatarsus varus are seen It is also termed metatarsus adductus Attention was called to this condition by Kite, and it is known by some as a third of a club foot. This colloquialism emphasizes that the deformity is in one plane only with the forefoot occupying an adducted position in relation to the hind foot. There appears to be no sex variation The condition may be unilateral or bilateral and there is often a tendency to turn the opposite foot out

Clinical Picture

The deformity is present at birth. An important clinical distinction should be made In some the forefoot can readily be pulled to a normal position aligned with the hindfoot by passive motion. In a few of these the baby's foot can be stimulated to active contraction of the everting muscles causing partial correction of the foot. The baby's foot tends to follow the stimulating finger stroking the foot rather than pulling away from it. The second and less common



Figure 21 The patient's right forefoot deviates medially forming a metatarsus varus deformity The left foot is carried medially by internal torsion of the tibia while the knee points forward.



Figure 22 Forefoot Varus, heel in neutral position in metatarsus varus deformity

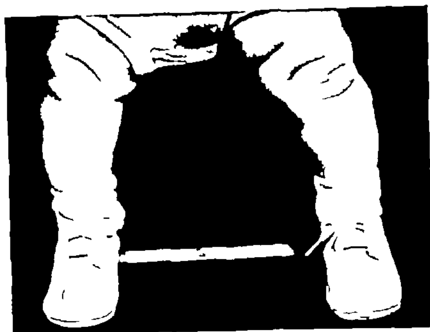


Figure 23 A bar holding the feet forward tending to correct internal torsion of the tibia and holding a corrected metatarsus varus deformity in straight alignment.

group cannot be passively corrected and constitutes a true deformity It is important to make this distinction as a basis for treatment.

There is usually an accompanying internal torsion of the tibia accentuating the turned in appearance of the foot. The patients often make their first appearance for treatment at the age of two or three months At that time the foot has become troublesome because it has failed to improve. This is due either to the patient s sleeping on his abdomen with his feet turned in (in the case of a mobile foot) or to the foot s being of a fixed variety

Treatment

MOBILE FOOT

Since it is not desirable to change the baby s sleeping habits the simplest method of getting the foot in good position while sleeping consists of using reversed shoes on a Dennis-Browne bar The shoes are placed on the bar in about forty five degrees of external rotation While the baby's awake reversed shoes or forefoot abduction shoes without the bar are used to avoid a tendency to develop genu valgus

In addition at each diaper change the mother holds the hindfoot and carries the forefoot into abduction with the other hand. She thus lines the foot up in normal position and stretches the structures of the medial side of the foot The usual time to procure correction is four to six months. When walking starts, straight last shoes are used as there is a tendency to recurrence.

THE FIXED FOOT

Here the metatarsus varus deformity must be stretched sufficiently to convert the foot to a mobile foot. This is best accomplished by corrective plaster

casts bringing the foot into full overcorrection. Either wedging casts or complete plaster changes may be used. Once the foot is mobile, the Dennis Browne bar with reversed shoes is used. Again this must be worn for a period of months or until the patient is walking. Shoes with a last carrying the forefoot into abduction are then used.

There is a definite tendency to recur which can be noted particularly when standing and indicates the necessity for prolonged follow up.

OVERLAPPING FIFTH TOES

The fifth toe, when structurally overlapped, has a tight skin fold and shortened tendon holding it in its place. This deformity is frequently familial. The condition is often bilateral and accompanying radial angulation of the distal phalanx of the fifth fingers is not uncommon.

Mild degrees of the condition may be strapped in place with adhesive over many months with the possibility of some success. When the toe is well up over the fourth, however, conservative methods will not correct the deformity. Surgical correction is indicated because of the tendency to develop a soft corn between the fourth and fifth toes and callus overlying the dorsum of the fifth. Correction includes a Z-plasty in the line of the short skin segment and lengthening of the extensor tendons and capsulotomy of the metatarso-phalangeal joint. At the conclusion of surgery the toe should lie naturally in normal position without having to be splinted.

CONGENITAL CLUB FOOT

Congenital club foot is one of the oldest known and treated deformities of mankind. Since the days of Hippocrates various methods of manipulation, strapping and binding have been used in its correction. The condition occurs once in every thousand births and is approximately twice as common in males. The characteristic deformity has three elements: forefoot adduction, inversion, and equinus of the foot. Such a foot has been termed talipes equino-varus. Although some reports have indicated an equal distribution between unilateral and bilateral types, most authorities indicate that the unilateral type is slightly more common.

Etiology

Duraswaimi produced this deformity in chick embryos after treating the developing embryo by insulin injections into its milieu in the egg. Despite this and other work indicating that perhaps lowered oxygen tension at a critical stage of limb bud development plays a part in its etiology, the cause of the deformity in humans is not established. Stewart notes a familial and racial predilection in his study of Hawaiian Island inhabitants. Bechtol and Mossman in dissecting two fetal specimens found evidence favoring muscle growth abnormality, resulting in failure to keep pace with skeletal development. The relative shortening of the muscle fibers was felt to be due to degeneration. Flinchum in studying a 6½ month premature infant, found the peroneal muscles on the involved side to be about one-half the size of the normal side.



Figure 24 Fifth toes overlapping bilaterally A fixed congenital deformity

This suggested an imbalance between medial and lateral musculature. Stewart in his dissections of club feet found abnormal insertions of the anterior tibial tendon apparently playing a part in producing adduction and supination of the forefoot. Other authors have emphasized abnormalities of the ligaments, (Brockman) and of nerves (Moore) The older German authors discussed bony anomaly—all pointing perhaps to the club foot being a faulty and incomplete development involving all structures controlling the position of the foot.

Clinical Picture

The orthopedist hopes to see a patient with a foot in an equino-varus position immediately after birth The immediate problem is to differentiate the positional foot from the fixed club foot The club foot cannot be passively corrected Stimulation of the peroneal muscles by stroking the lateral side of the foot reveals a deficiency in their action and their inability to evert the foot. Should the foot be mobile the expectation is that the foot can be readily and easily corrected. True club foot deformities are fixed The foot has fixed forefoot adduction fixed inversion particularly noted in the hindfoot, and equinus of the hindfoot carrying the forefoot with it.

The calf size should be noted. Smallness of the calf may be noted at birth before treatment is begun It tends particularly to be associated with the more rigid types of club foot such as are seen in arthrogryposis.

The degree of internal or external torsion of the tibia should be noted Surprisingly perhaps when the relation of the medial to the lateral malleolus is accurately determined it is found that they lie in the position of external torsion in relation to the knee. The medial deviation of the foot apparently is accounted for by the forefoot adduction.

The hips must be carefully checked for associated dysplasia or frank dislocation as is often seen in arthrogryposis. The opposite foot may be the site of less severe deformity such as metatarsus varus.



Figure 25 Plantar view of equino varus deformity revealing forefoot adduction, hindfoot inversion and equinus.

Figure 26 Club Feet. Posterior view



Treatment

Forcible methods of manipuation have been found in the past to be associated with the development of rigid insufficiently corrected feet This method was particularly done by means of the Thomas wrench—an instrument still found in the hospital instrument cupboard but now seldom used Kite has been responsible for the emphasis on gentle correction and an insistence on fully correcting one portion of the deformity before continuing treatment to other portions. If fundamental principles are followed most feet can be corrected into pliable, fully useful feet. About ten per cent or less remain which prove very resistant and for which other means may be indicated

Plaster casts in various forms constructed either in sections or as a complete unit may be used Others prefer the use of Dennis-Browne splints Whatever method is used the results should be carefully assessed both clinically and by roentgenogram to insure that full correction of all elements is obtained.

In Kite's method a plaster slipper is made correcting forefoot adduction.



Figure 9. Roentgenogram of uncorrected equino-varus feet with forefoot adduction and fixed equinu

Inversion is corrected by everting the slipper finally after the first two elements have been corrected the foot is brought out of equinus Such plasters are carefully applied without padding the foot to the point of obscuring land marks and the true position of the foot

Wedgings can be used but greater efficiency is obtained by complete changes of cast at weekly intervals Others use a cast applied as a unit to foot and calf The cast is then lengthened to a full leg cast holding the knee approximately in right angle flexion The heel must be well controlled to avoid slipping up in the cast This is avoided by molding the cast in well just above the prominence of the os calcis and the use of a full leg cast.

As Kite has emphasized holding the foot too long in a valgus position, before dorsiflexion is attempted will result in the creation of a flat foot. Pressure is not exerted on the forefoot to bring the foot into dorsiflexion This too fre-

equinus of the os calcis. A release of the medial ligamentous structures, capsulotomy of the ankle joint and heel cord lengthening or transfer of the medial portion of the achilles tendon to the lateral side of the os calcis is the usual procedure performed. Although known by various names it is most commonly termed an Ober ligamentous procedure

A wire through the superior posterior portion of the calcaneus may be added which is then used to control the position of the calcaneus as the plaster is applied and incorporated in the plaster The wire is removed at three weeks the plaster at six.

Follow Up Therapy

Feet that are fully corrected can be held thereafter in bivalved casts or forefoot abduction shoes attached to a Dennis Browne bar The mother also performs eversion and dorsiflexion exercises at each diaper change. She should be carefully instructed not to dorsiflex the forefoot, but let it be carried by the dorsiflexion of the hindfoot. The forefoot is stretched into abduction daily however

The patient is seen regularly by the orthopedist during the first two or three years and if the foot begins to lose mobility it is promptly placed in long leg plaster casts and wedged out into full correction. Short leg casts cannot fully control the forefoot adduction or the equinus

Anterior Tibial Transplant

Recurrence of deformity in apparently well-corrected feet may be due to peroneal weakness or imbalance between the anterior and posterior tibial and the peroneals

As emphasized by Garceau and Manning, if supination occurs on active dorsiflexion an anterior tibial transplant may be necessary to prevent recurrence of the deformity The usual age period when the need for transplantation becomes apparent is four to six years. The foot should be corrected prior to transplantation by wedging casts

The anterior tibial tendon when transplanted to the lateral side of the foot (base of the fifth metatarsal) has been found to be a weak dorsiflexor and to function poorly The usual procedure is to transplant the tendon to the base of the second metatarsal This position preserves the action of the anterior tibial as a dorsiflexor while weakening it as a supinator

The anterior tibial tendon is detached from its insertion and drawn up through a second incision into the calf It is then re-routed into the common extensor sheath and brought out on the dorsum of the forefoot. Here it is attached to the base of the second metatarsal through a drill hole or trap door

The Rocker Bottom Foot

Rocker bottom foot is a product of treatment. The foot has for practical purposes been overstretched in the mid tarsal region. This results in a prominent cuboid area which becomes the point of weight bearing with resultant



Figure 31 Recurring club foot with dorsiflexion in inversion



Figure 32 Uncorrected club foot walking with hindfoot in inversion

callus and disability There are at least four ways this foot can be produced (1) pushing the forefoot up out of equinus before the hindfoot equinus is corrected (2) slipping of the foot up into the cast so that the forefoot must be pushed up (3) walking on a foot with uncorrected equinus (4) exercises by the mother and others which dorsiflex the forefoot.

This tendency in the foot is very difficult to correct and the foot must be



Figure 33 "Rocker bottom foot with reversal of the lateral arch and cuboid prominence

Figure 34 Uncorrected equinus with break at mid-foot and relative dorsiflexion of the forefoot in "rocker bottom" foot.



held in correction for a considerable period The forefoot is pushed back down into equinus until it is lined up with the hindfoot in the cast The foot is then brought up as a unit until corrected.

When the foot cannot be corrected because of persistent equinus of the hind foot, a heel cord lengthening and ankle capsulotomy posteriorly may be necessary It is usual also to insert a wire into the posterio-superior angle of the os calcis to control its position. The wire is incorporated in the cast.

At six weeks the cast is removed but still further holding in plaster may be necessary perhaps two or three months with cast changes at approximately three-week intervals

Recurring Club Feet Below the Age of Eight

Wedging casts are resorted to in this period to restore full correction These feet presumably once were fully correctable and readily tend to wedge out. The forefoot adduction is wedged across the mid tarsal area. Equinus is corrected by wedges at the level of the ankle joint and not below it.

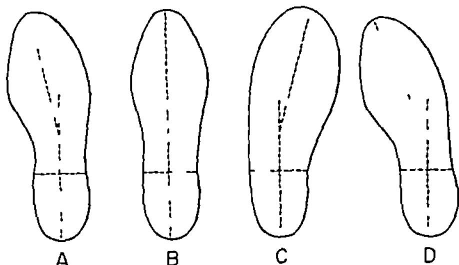


Figure 35 Varying types of shoe lasts controlling relation of hindfoot to forefoot. A regular B straight. C forefoot abduction. D forefoot adduction.

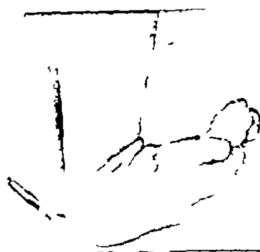


Figure 36 Calcaneo valgus foot with excessive dorsiflexion and tight anterior tibial.

Uncorrected Club Feet Over the Age of Eight

It is apparent that by this age considerable bony deformity has taken place in the feet A triple arthrodesis of the mid tarsal and subtalar joints can create a satisfactory foot for useful function without disability The Hoke arthrodesis with replacement of the astragular head is also useful Corrective wedges are removed so that the foot will lie in good position without forceful effort Such an operation is ordinarily not done before the age of twelve.

CALCANEO-VALGUS FEET

The calcaneo-valgus foot can be recognized at birth from its characteristic dorsiflexed and everted position.

The foot is very hypermobile except into equinus where it may be noted that the anterior tibial tendon becomes tight If the foot is fully mobile in all directions and actively corrects, it will tend to resume normal alignment in about two months

Some of these feet when checked by tight dorsal structures may be stretched out by corrective casts. The use of a Dennis-Browne splint with the cross bar inverted keeping the feet in that position has been suggested by Thomson. Such

treatment over a period of time sufficient to eliminate the excessive dorsiflexion and eversion is of value

Feet such as these tend to maintain their overstretched ligaments by the sleeping position When weight bearing begins the foot sags into severe pronation. A patient having had feet such as these at birth should be supported by stiff-soled shoes with one-eighth inch inner heel and sole wedges

CAVUS FEET

The characteristics of cavus feet—high arched feet with equinus of the forefoot cock up toes and a calcaneous position of the hindfoot—are readily recognized clinically This foot without too severe a deformity can produce severe disability They are very rarely seen below the age of three

Etiology

Such a foot brings to mind a triad of neurological possibilities as possible underlying diagnoses The first is an anomaly of the lumbosacral spine accompanying which may be adhesions interfering with root function. The resultant muscular imbalance is thought to result in the cavus foot A previous attack of poliomyelitis may result in muscular imbalance Weakness of dorsiflexors with posterior tibial still present may result in this situation Degenerative neurologic conditions such as Freidreich's ataxia may underlie this foot deformity It is important to recognize that chronic neurologic disturbance may exist and to take appropriate steps to diagnose it

Clinical Picture

These youngsters are usually recognized in the seven to fourteen year age group The changes in the foot by this time are well established The usual complaint is the deformity noted by the parents Symptoms in this age group are rare Calluses beneath the metatarsal head or over the proximal interphalangeal joint may be the presenting complaint

Suspicion of the foot deformity is aroused by equinus of the forefoot in non weight bearing. Confirming factors are the hyperextension deformity of the metatarsal phalangeal joints and flexion deformity of the interphalangeal joints resulting in claw toes These toes are subluxated dorsalward on the metatarsal heads The os calcis is not in equinus but in a calcaneous or dorsiflexed position giving rise to the high arch Passive elevation of the forefoot in this age group results in straightening of the toes.

Depending on the duration and severity of the deformity there may be thinning and atrophy of the fat pads over the metatarsal heads so that they may readily be palpated. Calluses may form over these bony processes The plantar fascia may be palpated as a tight contracted band on attempted correction of the forefoot and limit full correction

Treatment

Conservative treatment consists of passive stretching of the plantar fascia by dorsiflexion of the forefoot and flattening of the medial arch The toes are

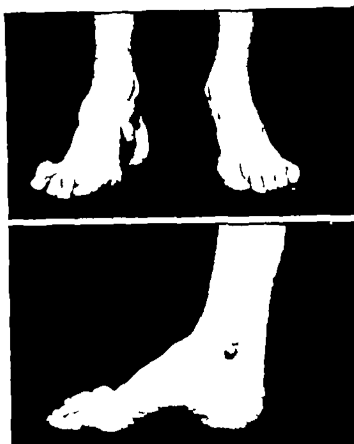


Figure 37 Cavus feet (note equinus of forefoot in relation to hindfoot)



Figure 38 Cavus foot

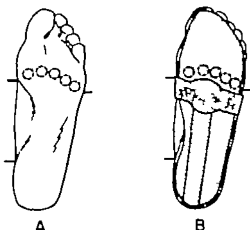
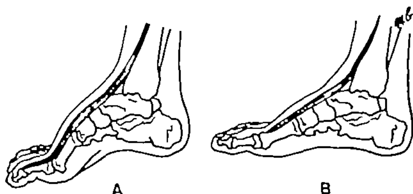


Figure 39 Tracing of pad with Jones bar to elevate forefoot.

Figure 40 Effect of moving long extensor of toes back to metatarsal neck correcting equinus of forefoot and allowing toes to drop down correcting dorsiflexion of the proximal phalanx.



brought down from their subluxated position on the dorsum of the metatarsal heads and the interphalangeal flexion deformity passively stretched.

A Jones Bar pad is either worn as an inner sole where it is most efficient or built into the sole of the shoe This pad to be corrective must place its elevation directly behind the metatarsal heads. In the still pliable foot this elevation restores the toes to their rightful position. The shoe aids in holding them down

Operation is considered in the presence of progressive deformity as a conservative regime and before the fat pads beneath the metatarsal heads have atrophied away

The operation that attacks the heart of the deformity is a transplantation of the long extensor tendons to the metatarsal necks. The short extensor tendons are detached at their insertion into the proximal phalanx and sewn into the stumps of the insertions of the long extensors. If necessary, a capsulotomy of the metatarsal phalangeal joints is done on the dorsum. An accessory bit of surgery sometimes necessary is a division of the plantar fascia. This is done just forward of its origin from the os calcis.

The most medial incision is used for the great toe Two tendons are approached through each of the other two incisions. The result of too few incisions is unnecessary retraction at the expense of vitality of the skin edges.



Figure 41 Detail of insertion of long extensor into metatarsal neck with insertion of short extensor into stump of longus.

A triple arthrodesis is sometimes used for correction of the deformity While this procedure corrects the deformity in part, it does so at only one series of joints, although many are involved, and it fails to attack the area causing disability—i e, metatarsal necks and toes Some correction in this area is achieved by the bony shortening relative to the soft tissues when a triple arthrodesis is done When this procedure is used, it is done in the older age groups but it has not proved as satisfactory as tendon transplant. After tendon transplant the foot is kept in a short leg plaster cast for six weeks

KOHLER'S DISEASE

Kohler's disease is a condition causing pain and swelling in the foot in which the roentgenogram reveals abnormal density of the navicular bone Involved in this way the navicular can be compressed if not protected Unfortunately this condition which is quite rare is often confused with the normal fragmentary ossification of the navicular which is not associated with symptoms

Normal ossification of the navicular from multiple centers occurs in the three- to six year-old age group Cases showing increased density of the navicular consistent with Kohler's disease usually occur past this stage in the six to ten year age group

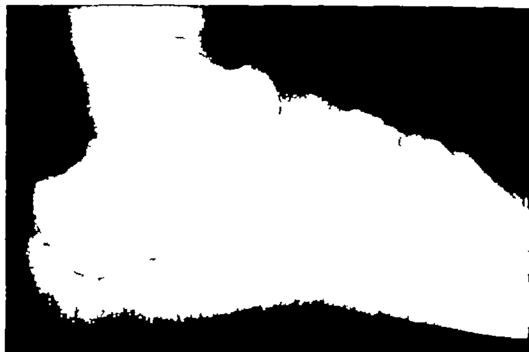


Figure 4. Roentgenogram of compressed and dense navicular in Kohler's disease with associated soft tissue swelling.

Etiology

This condition is usually thought of as secondary to circulatory interference. The reason for this change is not clear It is related presumably to other lesions such as Kienboch's disease Legg Perthes syndrome and Freiberg's disease of the second metatarsal head.

Clinical Picture

The patient complains of foot pain It tends to be continous rather than intermittent as in the case of foot strain There is tenderness located over the area occupied by the navicular bone Thickening and even frank swelling may be noted in this area. When the condition is active limp is noted with a tendency to walk on the lateral side of the foot

Roentgen Findings

The disease can be accurately diagnosed by roentgenogram. Confusion with normal ossification of the navicular can be avoided once it is realized that the navicular normally arises from at least three separate centers of ossification in the three- to six year age group These centers coalesce rapidly to form one center One overlying the other sometimes gives the appearance of increased density In Kohler's disease the navicular or a portion of it exhibits increased density and then fragments The radiolucent areas finally heal in to form normal appearing bone While going through the dense stage the navicular may become narrowed and compressed if the patient has been weight bearing.

Treatment

The patient is taken off weight bearing A protective plaster cast and crutches are used This is usually necessary for several months Before protected weight bearing with crutches is begun, the areas of density should be repaired and the soft tissue thickening relieved Full weight bearing is gradually achieved as the lesion heals

References

GARCEAU G J., AND MANNING, K. R. Transplantation of the anterior tibial tendon in the treatment of recurrent congenital club foot. J Bone & Joint Surg., 29: 1041 1047

HARRIS, R I AND BEATH T Hypermobile flat foot with short tendo-achilles. J Bone & Joint Surg 30A: 116 1948.

HARRIS R I., AND BEATH T Etiology of peroneal spastic flat foot. J Bone & Joint Surg 30B: 624 1948.

LAPIDUS P Spastic flat foot J Bone & Joint Surg., 28: 126 1946

THOMSON J Treatment of congenital flat foot. J Bone & Joint Surg 28: 787 1946

WHITMAN R. (Permission Lea and Febiger) A Treatise Orthopedic Surgery Ed 5 691 1917

2
The Knee

It is well to mention at the outset that the symptoms of knee pain in children may not be due to pathology centered at the knee. Hip-joint pathology frequently gives rise to knee pain. Foot strain may sometimes appear to give rise to knee symptoms

Lesions of the knee in children include entities such as osteochondritis dissecans discoid meniscus, and Osgood-Schlatter's disease. Popliteal cysts or Baker's cysts cause swelling posteriorly at the knee. Monarticular arthritis, a rheumatoid like lesion, occurs most commonly at the knee Foreign bodies sometimes give rise to a chronic swelling at the knee Septic arthritis and tuberculous synovitis are not uncommon.

OSTEOCHONDRITIS DISSECANS

Osteochondritis dissecans is known in adults but, in the presence of intermittent disability at the knee it must be considered in children. It is most common in males, and at least one-third of the cases are bilateral with a tendency to be symmetrical. The lateral aspect of the medial femoral condyle is the most common site although rarely it can occur in the lateral condyle and in the patella Osteochondritis dissecans occurs at the ankle hip and elbow but is so frequent at the knee that it is rarely thought of prior to roentgenogram at other joints. It has not been reported below the age of four

Pathology

From this point of view the lesion appears to be an aseptic necrosis of the subchondral bone for a variable but usually quite shallow depth. The cartilage is not primarily involved but becomes so when its underlying support is weakened It is possible to view the lesion with the articular cartilage intact and yet typical roentgenographic changes are present. The cartilage overlying the lesion secondarily develops fibrillation and cleaves from the cartilage areas which are well supported by normal subchondral bone. The fragment which separates includes subchondral bone and cartilage.

Etiology

It is assumed that the lesion develops because of localized interference with the blood supply to the area. The lesion has been likened to that which would

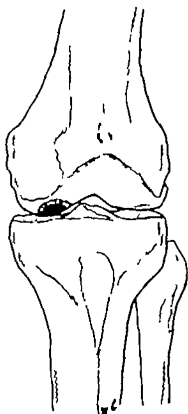


Figure 43 Diagram of characteristic site of osteochondritis dissecans slightly toward the lateral aspect of the medial femoral condyle

occur if a hollow sphere had been indented in a small area and that area then allowed to spring back into place. If the lesion is allowed to heal on a conservative regime it does so by creeping substitution The etiology is not clear, but trauma appears to be the initiating factor

Clinical Picture

Most characteristic perhaps is the intermittency of the symptoms Recurrent pain at the knee is the presenting complaint, with a further history of clicking giving way, and intermittent swelling at the knee. If a fragment has become detached, mild previous symptoms may be unnoticed in the acute and sudden onset of pain and swelling at the knee

Depending on the duration of symptoms a variable degree of atrophy of the thigh may occur Such atrophy reflects the disability that the patient has undergone Fluid may be noted in the knee but most diagnostic would be localized tenderness on the lateral aspect of the medial condyle. The intermittent symptoms may be the only reason for clinical attention with no signs of apparent disability

X Ray Findings

There is involvement of the subchondral bone A radio-lucent line separates a small dense fragment of this bone from the parent area. A minimal amount of reactive bone may occur about the lesion The lesion may not be clearly visualized in the usual anterior posterior view of the knee. A so-called "tunnel



Figure 44 Air injection of knee revealing defect in medial femoral condyle with fragment loose in center of the joint

view—taken postero anterior—with the knee flexed approximately thirty degrees is helpful in throwing the area most commonly involved into relief When the patella is involved a tangential view of the patella may be helpful

Treatment

Green and Banks have called attention to the fact that the lesion may be treated conservatively on a non weight bearing regime with good results. Such treatment using a non weight bearing splint or brace involves a healing period of approximately four to six months and perhaps longer The opportunity to heal the joint with intact articular surface makes this conservative regime the treatment of choice as compared to surgical excision which of necessity leaves a cartilage defect

Operative treatment has been frequently advocated and is followed by good results It has the advantage of a shorter period of convalescence—usually about

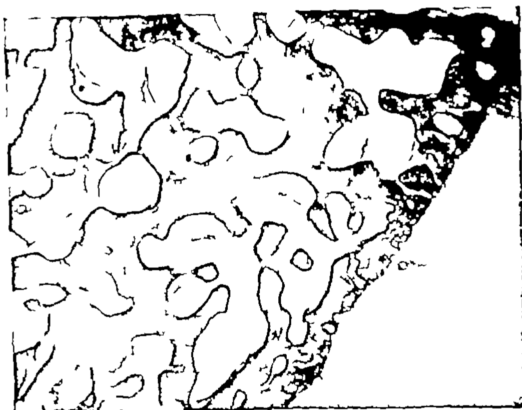


Figure 45 Photomicrograph of fragment from case shown in Figure 44 showing dead bone and articular cartilage

six weeks If the fragment of subchondral bone and cartilage has become detached its removal is indicated. When still partially attached the lesion when visualized can be eased readily from the surrounding bone A cleavage plane of fibrous tissue exists between the fragment and the parent bone.

When the cartilage is completely intact the area of subchondral weakness can be recognized by palpation and by a slight grayish discoloration. The lesion is cored out with smooth edges so that no unattached cartilage or bone remains. Drilling the base of the lesion may result in unwelcome exuberant repair and is not recommended.

CONGENITAL DISCOID MENISCUS

Congenital discoid meniscus is a lesion of the cartilagenous meniscus of the knee primarily occurring in the lateral meniscus

Normal Development

The fetal meniscus begins differentiating into the adult type structure at the fifth month The meniscus originally consists of a complete disc which then softens in its central portion A gap appears which has only partially progressed at the time of birth The gap gradually increases in size thereafter until the more adult form is reached The area of direct contact of the opposing articular surfaces is always greater on the medial than on the lateral side in the normal knee.



Figure 46 Discoid Lateral Meniscus (From Smillie I S Injuries of the Knee Joint. The Williams & Wilkins Co Baltimore Maryland 1951

Etiology

Smillie points out that a primitive disc has its greatest thickness in the central unabsorbed segment The normal resorption of the meniscus in its central portion has not taken place The central portion is subject to direct compression as a result a horizontal cleavage plane may appear between its upper and lower portion which when carried to its full development results in a hole in the cartilaginous disc

A feature in the course of this development is a ridge produced when the femoral condyle impresses into the disc as the inferior surface of the cartilage gives way These ridges aid in explaining the clinical symptoms. The disc itself is not thicker than normal.

Clinical Picture

Classically as emphasized in the literature the basic complaint is a snapping knee. It is often noted that the child s knee clicks in flexion and extension. The sign that makes the diagnosis as certain as it can be is a click elicited as the fem



Figure 5 Central defect in primitive disc produced by horizontal cleavage (From Smillie I S Injuries of the Knee Joint. The Williams & Wilkins Co Baltimore Maryland 1951

oral condyle alters its relation to the disc by jumping over the ridge and pushing the cartilage either forward or backward as the case may be The impingement of the cartilage by the femur and tibia in its central portion fixes the disc, so that of necessity the anterior and posterior attachments are stretched. Signs not elicited when the child is young may gradually become more apparent as the disc becomes more loosely attached. Older children may complain of giving way of the knee There may be episodes of swelling and fluid.

Treatment

Excision of the disc on the basis of the abnormal mechanics at the knee which it introduces is justified. The recovery period following such excision is greatly shortened if the youngster has been taught quadriceps-setting exercises prior to

surgery Such exercises are difficult to teach the child when he has a painful knee in the immediate postoperative period. Crutch walking is begun when the foot can be lifted from the bed with the knee extended Full activity is allowed in the presence of full motion a strong quadriceps and an absence of swelling at the knee. The approximate period of convalescence in a child is six weeks.

OSGOOD-SCHLATTER'S DISEASE

Osgood-Schlatter's disease a disease of adolescence causes pain anteriorly at the knee. Osgood first called attention to the enlargement of the tibial apophysis in 1903 The entity is best understood when thought of as a tendinosis rather than an apophysitis however Hughes notes that the common feature by roentgenogram is calcification within or enlargement of the ligamentum patellae. The disease is self limited, running a two- to three-year course The age of incidence runs from ten to sixteen years It is slightly more common in males and unilateral cases occur twice as often as bilateral.

Etiology

A history of an extremely rapid growth spurt in the year preceding onset of symptoms is readily obtained. One suspects that the increased soft tissue tension thereby produced causes abnormal stress at the apophysis of the tibia. Such stress is directly transmitted by the ligamentum patellae There is resultant swelling of the tendon at its insertion and secondarily irregular ossification Changes very similar to Osgood-Schlatter's disease can be produced in the rat by shortening the patella tendon The disease in man ceases at the time of fusion of the apophysis to the parent bone if it is still active at that time Growth plays an important part in its etiology by producing abnormal stress at the tibial apophysis It is possible that trauma to the tibial tubercle may also produce such stress

Clinical Picture

The history reveals that the discomfort is experienced with activity and decreases with rest Sports which involve much running and jumping are particularly likely to produce exacerbations of the disease. A rapid growth spurt is common in the year preceding onset. Discomfort and swelling are located anteriorly in the region of the tibial apophysis. There is no swelling involving the joint itself The thickening and enlargement of the tibial tubercle is characteristic.

Careful palpation reveals that tenderness is located at the point of insertion of the tendon into the bone but not over the apophysis itself Extreme flexion is painful Atrophy of the quadriceps is not remarkable unless the symptoms have been severe

Roentgenogram

The apophysis ordinarily appears at the age of eleven, and fuses with the parent epiphysis at fifteen in males It is most hazardous to base the diagnosis on the presence of irregular ossification of the apophysis. Such ossification forms may be part of normal development. The one constant finding is swelling of the tendon shadow at the area of its insertion into the bone. The swelling frequently



Figure 48 Swelling of the area of patellar tendon insertion into the tibial tubercle in case of Osgood Schlatter's Disease

obliterates the angle formed between the tendon and the bone. In some cases adventitious ossification may proceed into the tendon itself.

Treatment

The discrepancy in functional length between soft tissue and bone may take two to three years to adjust. When there is no longer abnormal soft tissue stress, the symptoms may cease even before fusion of the apophysis takes place. When the symptoms are severe rest is enforced by strapping with adhesive. If necessary a plaster cast in the form of a walking cylinder may be applied.

Limitation of activities voluntarily is often possible in this age group, particularly when the relationship between activity and symptoms is pointed out to the patient and his parents. The patient can still be ambulatory, but should resist sports running and long walks. The relationship between activity and symptoms is a very direct one and readily appreciated by the patient once it is pointed out. The limitation of activity is usually necessary only for a period of approximately six months. Once the functional length of the soft tissues has caught up with the bone full activity can be resorted to by the patient with impunity.



Figure 40 Swelling of patellar tendon insertion and fragmentation of the tibial apophysis by roentgenogram.

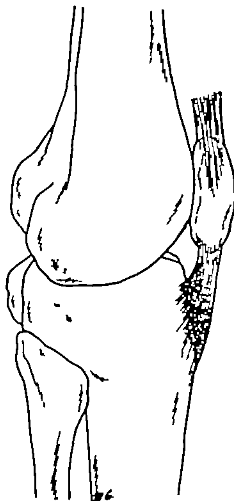


Figure 60 Diagram of the area of swelling in Osgood's-Schlatter's Disease.

The thickening and enlargement of the apophysis persists even after the disease heals and symptoms subside. The enlargement itself may be a source of discomfort in kneeling. This discomfort persists after the disease is no longer active.

POPLITEAL CYSTS

Popliteal cysts have also been designated "Baker's cysts." Baker in 1877 described cases of swelling in the popliteal space and thought that perhaps they were herniations of the knee joint space which had been chronically distended by fluid. Faucher in 1856, had described eleven dissected specimens under the impression that they represented distension of bursae located in the region of the medial head of the gastrocnemius-semimembranous bursa which had become filled with fluid as a result of chronic irritation

These cysts are not uncommon in children about one-third of the cases occurring in the one- to fifteen year age group They are most common in males (by a ratio of two to one) and are ordinarily unilateral. The etiology of these cysts in adults may be quite different from that in children Most children have a history of no previous effusion or symptoms at the knee when the cyst is discovered.

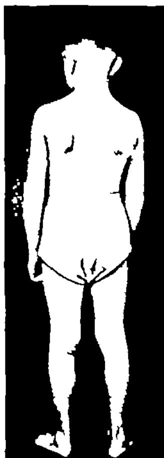


Figure 51 Popliteal cyst, posterior view

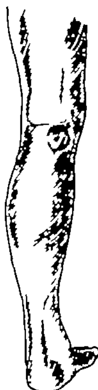


Figure 52 Diagram of popliteal cyst location—medial posterior aspect of knee and inferior to knee crease

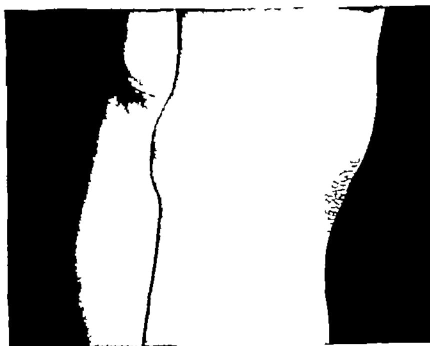


Figure 53 Popliteal cyst, lateral view

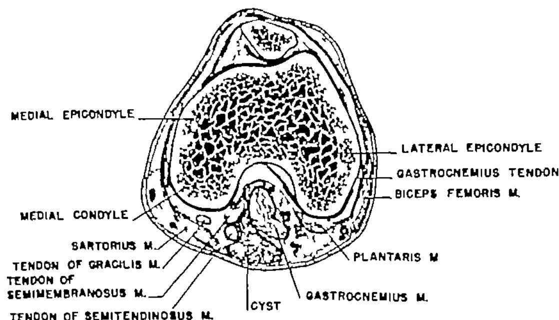


Figure 54 Anatomical diagram showing location of popliteal cyst in anatomical site of semimembranosus bursa (*Redrawn from* Wilson, P D Eyre-Brook A L and Francis, J D Clinical and anatomical study of semimembranosus bursa in relation to popliteal cyst. *J Bone & Joint Surg* **20**: 963 1938)

Clinical Picture

The complaint is that of the swelling itself in children There are no symptoms of the knee joint dysfunction. In the adult these cysts are often associated with osteoarthritis The symptoms referable to the knees are usually on the basis of the arthritis and these symptoms are due to the arthritis itself

The popliteal area is one that is very difficult to palpate and one may often be misled into believing that a cyst exists In general, when one does exist, there is no doubt. The swelling tends to lie distal to the knee crease and on the medial side of the posterior aspect of the knee

Roentgenogram

The X ray picture should be of help in confirming the clinical impression A smoothly outlined swelling of fluid density is present in the popliteal space This can be noted particularly in contrast with the picture of the other knee The fact that the cysts are so rarely bilateral is of great help

Treatment

Excision is recommended in the usual case Wilson *et al.* dissected the gastrocnemio-semimembranosus bursa in 1938 and pointed out the anatomic relationship of this bursa to the popliteal cyst. Invariably there is an intimate attachment of the bursa to the tendinous portion of the medial head of the gastrocnemius and semimembranosus muscles requiring a partial resection of these structures to maintain the cyst wall. The cysts often communicate with the knee joint capsule through an opening on the proximal portion of the medial condyle beneath the medial head of the gastrocnemius Wilson states that this

connection was found in fifteen of twenty-one operated cysts and in seventeen of thirty anatomic specimens of the bursa.

Examination of the cyst walls after removal reveals fibrous thickening, with areas of retained hemosiderin and areas of metaplasia into cartilaginous and osteoid elements.

MONARTICULAR ARTHRITIS

Monarticular arthritis consists of chronic synovial thickening with joint fluid findings similar to rheumatoid arthritis, but limited to one joint. It may occur at other areas such as the ankle, hip, and elbow, but is so characteristic at the knee that it is here fully described. The age group is not well defined, but most cases occur between five and ten years of age. The disease is slightly more common in females.

Etiology

The etiology is very closely related to rheumatoid arthritis—so closely that *some cases under observation will be noted to develop other involvement.* Imperceptibly, thickening and fluid develop at other joints and obtain the more typical rheumatoid distribution. Aspiration of fluid from the joint reveals cellular, chemical changes similar to those found in rheumatoid arthritis. Nonetheless this entity is very frequently limited to one joint and remains so. Amelioration of symptoms is possible with intelligent care.

Clinical Picture

The child presents chronic swelling of the knee often of several weeks or months duration. Pain is not a prominent feature but a limp is usually noted by the parents. The knee reveals synovial thickening with fluid filling the suprapatellar pouch which produces a ballottable patella and fluid wave in examination. There is increased heat and moderate tenderness of the anatomic area occupied by the joint. The knee lacks ten to twenty degrees of full extension. Full flexion is also limited. There is palpable and measurable atrophy of the quadriceps.

Laboratory Findings

Laboratory work reveals an increased sedimentation rate and occasionally a mild leukocytosis. Aspiration of the joint may reveal a considerable elevation of polymorphonuclear cells and the total cell count may reach 3,000 to 4,000 cells per cubic millimeter. The joint sugar may be depressed in relation to the blood sugar taken at the same time—that is, by more than twenty milligrams per cent. A positive tuberculin test (in the absence of a clear-cut clinical differentiation from tuberculosis) may necessitate biopsy of the joint to establish the diagnosis.

Treatment

The child often comes in walking with a knee-flexion contracture. This position of strain in weight bearing will not help a low-grade condition of this type





Figure 55 (left) Lateral and anterior view of knee in monarticular arthritis. There is thickening of the capsular shadows about the joint and posterior subluxation of the tibia



Figure 56 (right) Knee flexion contracture in ambulant child



Figure 57 Swelling of the suprapatellar area, due to synovial thickening and fluid can be seen

to clear Traction is often advisable to secure full extension. Bivalved plaster casts may be used with non weight bearing exercises to secure the same result

Rest is still the therapy of choice for monarticular arthritis although hydrocortone injections into the joint may be of some help The absolute rest from weight bearing is achieved by means of a bivalved cast or removable splint holding the knee just off full extension.

It is important that the parents understand the chronicity of the lesion. The child is usually at rest for four months, with a very gradual resumption of weight bearing (via crutches) over the following two months. Signs of recurrence of fluid are regularly sought and the sedimentation rate checked.

The joint handled tenderly and slowly responds first by losing its fluid and thereafter by a gain in the range of motion loss of heat and subsidence of most of the synovial thickening. Some slight thickening may remain. Rest during the early months includes complete bed rest and a home teacher should be arranged for if the patient is of school age.

A careful and gradual resumption of activity for the joint is usually rewarded by a complete cure except for those cases which pass gradually over into rheumatoid arthritis.

THE FORIEGN-BODY KNEE

A picture very similar to monarticular arthritis is sometimes seen in a patient with a definite history of trauma in the months immediately preceding consul

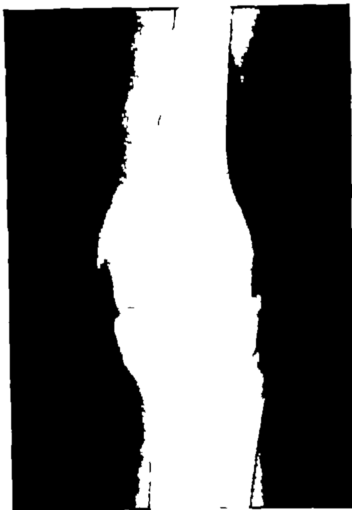


Figure 58 Air arthrogram of knee with pieces of glass present laterally and in the femoral notch

tation. The trauma is such that pieces of glass wood cinders or clothing could have been driven into the knee

The patient has a unilateral chronic swollen knee X ray may reveal small bits of foreign body but often does not. The joint fluid often contains an unusual amount of fibrin clot

Incision into the joint reveals a large fibrin clot filling the suprapatellar pouch This knee may require synovectomy of the accessible portions of the joint to effect a cure The synovectomy must be followed by traction and physiotherapy, including range-of motion exercises and active exercises of the quadriceps musculature to effect a good result Synovectomy should not be undertaken until it has been demonstrated that removal of the fibrin clot and accessible foreign material will not effect a cure

SEPTIC ARTHRITIS OF THE KNEE

Septic arthritis of the knee is not as common as involvement of the hip because the metaphyseal area is not enclosed within the joint.

Etiology

When acute it is most often blood borne—a complication of a severe childhood septicemia. Below the age of one the most likely etiologic agent is the streptococcus, the older age group is more often involved with the staphylococcus Direct involvement of the knee by foreign bodies following puncture wounds gives rise to a low grade type of septic arthritis, which is frequently a puzzling diagnostic problem

Clinical Picture

The acute septic knee gives rise to apprehension pain at the knee and the signs of sepsis elevated temperature and increased pulse The swollen joint is extremely tender to palpation. The knee is held in a flexed position with marked hamstring spasm. Swelling is most apparent in the region of the suprapatellar pouch.

Joint Fluid Findings

Aspiration reveals a cloudy white to thick purulent fluid. Cells are usually over 4 000 per cubic millimeter and consist almost entirely of polymorphonuclears. Smear with a Gram stain frequently reveals the organism

The fluid clots on standing it contains increased protein and frequently clumps of fibrin. The *joint sugar is depressed more than twenty milligrams per cent below the blood sugar taken at the same time*

Aspiration is performed from the lateral side into the suprapatellar pouch just proximal to the upper pole of the patella A needle such as is used for spinal *puncture is most valuable—at least nineteen gauge in size and preferably eighteen gauge* The aspiration is likely to be more successful if pressure is exerted on the joint and the fluid milked out rather than withdrawn by suction. *Chemotherapeutic agents in appropriate concentration may be inserted into the joint following aspiration. Cultures made of joint fluid should* be both anaerobic and aerobic In a small percentage of joints the suprapatellar bursa does not communicate with the joint space and aspiration must be done into the joint space on either side of the patella at its lower pole

Treatment

The patient is made comfortable by traction in line with the flexion deformity of the knee This is most simply done by using both an under the-knee sling and a *foot strap this is called Split Russell traction*

Following aspiration chemotherapy is instituted in line with that indicated for the suspected organism. As a rule the symptoms tend to subside rapidly Intra articular insertion of the chemotherapeutic agent may be continued In *some cases the suprapatellar pouch and joint remains thick and boggy* The temperature continues to hover at a slight elevation the sedimentation rate remains elevated It may be necessary to drain such a joint if symptoms do not rapidly subside Incision into the joint reveals it to be filled with fibrin clot. *Once the joint is emptied the synovia is* repaired with plain catgut. A catheter

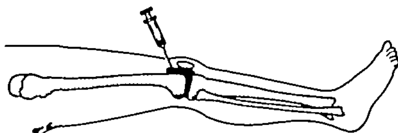


Figure 59 Aspiration of knee from supra patellar pouch In a small percentage of cases the pouch does not communicate with the knee joint itself

for the instillation of appropriate chemotherapy may be left in place, but the remainder of the wound is repaired *per primam* The expected course is that of subsidence of symptoms

TUBERCULOSIS OF THE KNEE

Tuberculosis at the knee in childhood occurs in a very typical form which must be differentiated from other causes of chronic joint effusion.

Etiology

The disease is metastatic to the knee following primary involvement of the lung and is due to the tubercle bacillus originally described by Koch in 1882

Pathology

It is most common for the synovia to be initially involved. Rarely the epiphysis of either the femur or tibia is first involved, followed by a secondary involvement of the synovia. In the typical early case the synovia is markedly redundant, tending to fill the joint space. There is an increased amount of joint fluid. The synovia tends to have a grayish discoloration but when viewed at operation under tourniquet appears plum colored. Tubercle formation is usually not visualized in the gross but is demonstrated microscopically The articular cartilage appears uninvolved although this appearance may be deceptive. The portions that are involved are usually covered with adherent synovial membrane

In late cases there is extensive involvement of the ligamentous structures. The result is great disorganization of the joint Such progression of the disease leaves little hope for preservation of the joint as a functioning structure. There is ordinarily a past history of exposure to an individual suffering from tuberculosis or a record of previous involvement which may include tuberculous glands or meningitis as well as disease of the lungs.

Clinical Picture

It is usual to see the patient after swelling of the knee has been noted for several months Pain is not a prominent feature in the synovial tuberculosis seen at the knee

Palpation reveals a plastic type of protuberance in the suprapatellar region



Figure 00 Synovial tuberculosis of the knee The normal anatomical outlines proximal to the patella have been obliterated by swelling of the suprapatellar pouch on the right.

Areas where the synovia can be palpated are thickened and tender A fluid wave can often be demonstrated There is usually a flexion contracture of the joint.

The tuberculin test is positive Where the disease is suspected it is best to begin testing with a dilution of 1 100 000 The sedimentation rate is elevated and the white blood cell count though not markedly elevated frequently reveals a lymphocytes to-monocytes ratio of well over five or six to one

Aspiration of the joint reveals white cells ranging around 1,000 to 2,000 per cubic millimeter with a preponderance of polymorphonuclears The fluid is slightly cloudy to turbid and may contain flecks of fibrin. The joint sugar is usually more than twenty milligrams per cent lower than a blood sugar taken at the same time.

Roentgenogram

There is a shadow of water density filling in the suprapatellar pouch area and rounding out the normal anatomic shadows of the joint. Signs of chronicity are apparent in the demineralized bone In cases of fairly long duration the joint space is narrow Rarely an area of bone destruction may be seen in the epiphysis.

Treatment

The absolute rest enforced on tuberculous patients with regard to their joints from the days when the disease was first recognized remains a dominant theme in treatment to the present day Joint fusion was developed to aid in obtaining

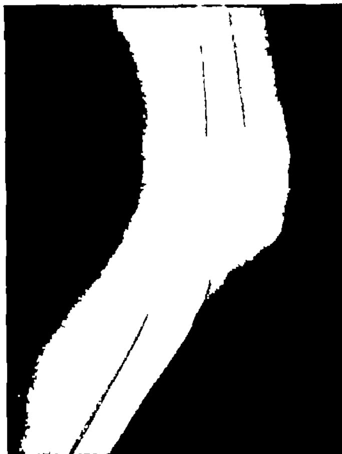


Figure 61 Thickened capsular shadows and filling of suprapatellar pouch area with shadows of soft tissue density in synovial tuberculosis of the knee

complete rest and the healing of the disease Such fusions saved the epiphyseal discs, but in children are subject to progressive deformity at the knee The soft bone in this area allows considerable flexion to develop

A unilateral hip spica is necessary to secure rest for the knee and should be applied after biopsy is performed. Biopsy is usually necessary to confirm the diagnosis. The present-day tendency is to try to preserve knee motion under the cover of the chemotherapy of tuberculosis There is no reason to abandon rest as a principle, however and to do so leads to recurrences of the disease.

The spica is bivalved and affixed with canvas straps, from this the leg is removed twice daily for exercise. The usual duration of treatment with chemotherapy and spica is seven months for synovial disease of the knee Synovectomy of the knee results in considerable impairment of motion and in view of the excellent possibility of obtaining healing of tuberculosis of the knee in childhood is not thought necessary

PARA AMINO-SALICYLIC ACID

This drug is inadequate when used alone Resistance of the organism develops ordinarily after six months of its use Its greatest value is in the prevention of emergence of strains resistant to streptomycin when used in conjunction with that drug.



Figure 63 Synovial Tuberculosis of the knee The joint surface is at the top with tubercle formation, mononuclear phagocytes, lymphocytes and giant cells below

Streptomycin

This drug has been found as effective in divided doses twice weekly, as it is when given daily When given daily and alone streptomycin has been shown to produce resistant strains after 42 days in 35 per cent after 60 days in 50 per cent and in 75 per cent after 120 days.

The effect of streptomycin on the eighth nerve may leave permanent deafness and troublesome vertigo

Isonicotinic-Acid-Hydrazide

The use of this drug with streptomycin will delay the development of resistance which occurs with greater rapidity when either is used alone. Isonicotinic-

acid hydrazide, when used singly, is followed by the development of resistant strains in the second three months of treatment. This drug apparently has the ability to penetrate cells and affect the tubercle bacillus There is a very definite synergistic effect with streptomycin and the bactericidal property of both is enhanced when used together

Para amino-salicylic acid, isonicotinic acid hydrazide and streptomycin are apparently more effective when used together since the drugs are usually used for periods longer than three months Persistent strains tend to die out when streptomycin is discontinued, and second courses may be given with effect.

Chemotherapy allows a period of conservative therapy to be shortened and results in a greater likelihood of obtaining a cure and a functioning joint. Traction can be used as an adjunct to rest in a bivalved spica to obtain motion at the knee once the disease is controlled There should be a normal sedimentation rate and a restoration of a normal lymphocyte-monocyte ratio The resumption of weight bearing should be gradual, and delayed for a six month period after chemotherapy and the cessation of signs of activity of the disease During this period the patient may walk in a non weight bearing splint. From this point progress may be made in the absence of signs of recurrence to partial and, finally, full weight bearing. The patient should have ninety degrees of flexion, full extension, and a good quadriceps before weight bearing is resumed.

CONGENITAL GENU RECURVATUM

Congenital genu recurvatum is a striking deformity at birth The tibia is markedly hyper-extended on the femur with the skin wrinkled and creased anteriorly and stretched tightly posteriorly Despite the apparent severity of the deformity on observation, the outlook is good. The underlying etiology appears to be intrauterine position, the knee appearing normal in other respects. A history of breech birth is not unusual and accompanying dislocation of the hip is not infrequent.

Treatment consists of the repeated application of corrective plaster casts. The knee is gradually brought back to neutral and then,flexed The knee must be held in flexion for a growth period of several months, once corrected. Such a period allows reestablishment of normal muscle lengths relative to the bones.

A tendency to recurrence indicates the need for bivalved casts and exercises before walking and a period of bracing may be necessary when the child begins to walk. Actual dislocation, if present, in addition to hyperextension may demand more radical treatment to get the tibia beneath the femur in normal position.

RECURRENT DISLOCATION OF THE PATELLA

A good illustration of the effect of body mechanics and growth in the production of disease is contained in the entity known as recurrent dislocation of the patella. The patella may be dislocated at birth and remain in this position unless surgically corrected. Simple dislocation may occur due to trauma. Recurrent dislocation is a result of an abnormal direction of pull of the quadriceps muscle and may or may not be accompanied by other structural abnormalities which

render dislocation easy of accomplishment. It is twice as common in females and tends to be familial.

Etiology

In order to produce a recurrent dislocation the quadriceps musculature—which contains the patella in the manner of a sesamoid bone—must, when contracted tend to pull the patella laterally The factors in the development of this lateral force rather that a force running straight through the condyles of the femur include structural abnormalities which carry the patella tendon insertion laterally This is true in genu valgus and in external rotation of the tibia. Both of these structural entities may result from abnormal growth secondary to unequal muscle pull, as in poliomyelitis or from congenital anomalies of the lower legs.

An absence of the vastus medialus from whatever cause, underdevelopment of the lateral condyle of the tibia, failure of the intercondylar groove to form—these all aid in the production of dislocation. An unusually long patella tendon allowing the patella to lie above the intercondylar groove also cases lateral displacement.

Mechanism

For functional purposes the quadriceps musculature may be visualized as contracting in a straight line between the origin of the rectus femoris or the anterior inferior spine of the ilium and its insertion via the patella tendon into the tibial tubercle or apophysis. This line should run directly between the condyles of the femurs in the intercondylar groove Either valgus or external rotation of the tibia in relation to the knee may carry the tibial tubercle lateralward. When the patient is unusually long limbed a small lateral displacement has a greater effect toward producing lateral motion of the patella.

The lateral condyle of the femur ordinarily provides a bony hindrance to lateral movement of the patella The vastus medialis and medial capsule provide a ligamentous hindrance Underdevelopment of the lateral condyle paralysis or absence of the vastus medialis and laxity of the medial capsule eliminates these check reins. Any all or none of these features may be present.

A patella tendon allowing the patella to ride above the intercondylar notch eliminates the lateral condyle as a buttress in preventing lateral dislocation and may be a contributing mechanism.

Clinical Picture

The child is usually over the age of six and tends to be in the early adolescent group A period of rapid extremity growth often precedes the development of the condition. The rather tall long legged early adolescent female with genu valgus of moderate severity is the typical subject.

The dislocation is usually not seen. The child is brought to the physician because of recurrent pain or swelling at the knee. The actual dislocation which occurs when the knee is flexed ordinarily reduces itself when the knee is extended. The history usually contains several episodes of difficulty The most likely time

A B C

Figure 63 Two factors affecting the line of quadriceps pull. A normal B valgus at the knee C external rotation of the tibia. Lateral displacement of the patella tendon insertion tends to carry the patella laterally on contraction of the quadriceps.



Figure 64 Dislocated patella in six-year-old girl. The apparent patellar prominence is due to a fat pad. The actual patella prominence can be seen above and lateral to it.

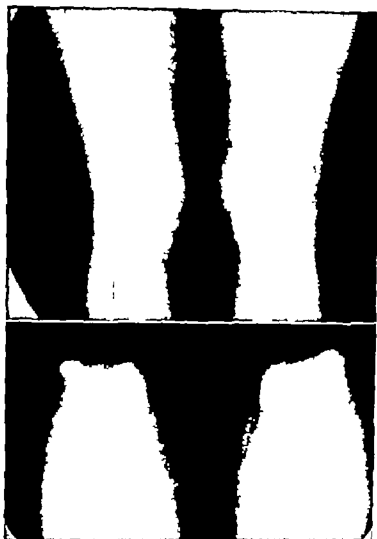


Figure 65 Roentgenogram of bilateral dislocated patellae Antero-posterior view above and below view with ray directed tangential to the patellae as the knee is flexed.

of occurrence is following periods of prolonged sitting such as would occur when the child is entertained On arising the inability to straighten the knee is noted, and the displacement of the patella may actually be appreciated although often it is not.

When seen within forty-eight hours of the dislocation, the knee usually contains fluid. The medial capsule of the knee in the parapatellar area is tender Atrophy of the thigh is variable dependent on the degree of previous disability

A helpful sign in arriving at a diagnosis whether the child is seen immediately after the dislocation or many months later is the acute apprehension on attempted lateral displacement of the patella When the knee is extended and the quadriceps relaxed passively pushing the patella over the lateral condyle of the femur will give rise to acute apprehension as the patient feels the patella about to slip out of control

Structural abnormalities are noted. A mild valgus position of the tibia will result in considerable lateral displacement of the tibial tubercle in a tall, long limbed child. There may be external rotation of the tibia, again with lateral displacement of the tibial tubercle.

Roentgenology

The antero-posterior view of the knee may reveal the dislocation of the patella, if present. The tendency for the reduced patella to ride more laterally than usual and valgus at the knee may be noted. A lateral film may reveal the patella to be proximal to or at the superior edge of the femoral condyles. A view taken tangential to the condyles to show the intercondylar notch and the relation of the patella to it may reveal the dislocation or the underdevelopment of the lateral condyles.

Treatment

The fundamental approach in treatment is to correct the line of pull of the quadriceps so that it runs through the intercondylar area. Since a lateral force is needed to produce the dislocation, it follows that correction of this force will result in a cure of the condition.

The reasons for attempting cure are alleviation of episodes of discomfort, production of normal function and protection of the undersurface of the patella from further injury. Chondromalacia patellas seen in later life often gives a history of recurrent or chronic dislocation of the patella in childhood.

The treatment may be complicated by the patient's age. Below the age of fourteen in girls and sixteen in boys interference with the growing apophysis forming the tibial tubercle may give rise to growth deformity. Over this age the mechanical line of pull of the quadriceps can readily be corrected by transference medially of the patellar tendon insertion. The age of election for surgery for any individual patient varies.

In the young age group, when the recurrent dislocations are frequent and productive of fluid in the knee, temporizing measures may have to be undertaken.

References

BAKER, W. M. On the formation of synovial cysts in the leg in connection with disease of the knee joint. *St. Barth Hosp Rep* **13:** 245 1877

BOSWORTH, D. M., PIETRA, A. DELLA, AND FARRELL, R. F. Streptomycin in tuberculosis bone and joint lesions with mixed infection and sinuses. *J Bone & Joint Surg* **32A:** 103 1950

HUGHES, F. S., MARDIS, R. E., DYE, W. E. AND TEMPEL, C. W. Combined intermittent Regimens in the treatment of non-miliary pulmonary tuberculosis. *Trans Tenth Conference Chemotherapy of Tuberculosis* p. 67 Washington Veteran's Administration—Army Navy

SMILLIE, I. S. The congenital discoid meniscus. *J Bone & Joint Surg* **30B:** 671 1948.

WILSON, P. D., EYRE BROOK, A. L., AND FRANCIS, J. D. A clinical and anatomical study of the semimembranosus bursa in relation to popliteal cyst. *J Bone & Joint Surg.*, **20:** 963 1938

3

The Hip

CONGENITAL HIP DISEASE

The very noticeable limp due to weakness of the hip abductors when the hip is unstable has been with mankind since the first civilizations Hippocrates (460–370 B C) wrote

> There are persons who from birth or from disease, have dislocations outward of both thighs in them then the bones are affected in a like manner but the fleshy parts in their case lose their strength less the legs, too are plump and fleshy except that there is some little deficiency at the inside and they are plump because they have the equal use of both legs, for in walking they totter equally to this side and that.

An enlightened medical community has succeeded in recent years in recognizing states which predispose to dislocation and thereby has greatly reduced its incidence Hart has emphasized the preventive nature of treatment undertaken *within the first year of life to form an adequate hip joint in an individual who* might otherwise go on to dislocation of a previously dysplastic hip joint. Hart has also brought out the end result of an uncorrected subluxated hip which may go on to degenerative arthritis in adult life

Dislocation may be present at birth but also may occur in the first years of *development. Dysplasia or pre-subluxation may be the first* indication of abnormal mechanical forces which may eventually result in dislocation. The work of Hart Hass and the work of the Italian orthopedic surgeons such as Putti, Scaglietti and Poli has been responsible for the recognition of these states other than frank dislocation.

Gill Crego and Schwartzmann and others have noted that the age when treatment started may be correlated with the excellence of the end results in terms of percentages. The number of failures rises as one progresses in to the older age groups.

The sex incidence of congenital dislocation in various reported series has varied from eighty-one per cent to eighty seven per cent in favor of females. Hass states that the female is affected six times as frequently as the male. Unilateral dislocations are twice as common as bilateral when a frank dislocation is spoken of *Various forms of hip dysplasia* when included particularly



Figure 66 Old uncorrected subluxation of the hip with degenerative changes

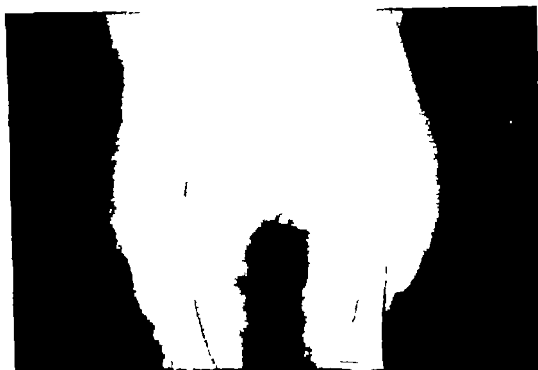


Figure 67 Unilateral dislocation of the hip Note increased acetabular slant, diminished size of the epiphyseal growth center lateral and superior position of the femur and anteversion as shown by the externally rotated position of the proximal femur in relation to the antero-posterior view of the distal femur

in the age group under one year, increase the incidence of bilateral involvement so that it is more nearly equal to that of unilateral difficulty

Congenital dislocation of the hips becomes extraordinarily frequent in some areas. There are local areas that have been reported in France Holland and Italy where the incidence of the disease appears to be greatly increased. The northern provinces of Italy are an example. It appears that the disease is exceptionally rare in the Negro In Asiatic countries, where the child is balanced on the mother's hip with the legs in abduction as a characteristic means of carrying the child the incidence of poor hip development is much reduced.

Congenital dislocation of the hip is a poor term for a lesion that often starts off as a slanting acetabular roof and underdevelopment of the proximal end of the femur, and progresses to dislocation as the result of mechanical forces acting upon it.

Etiology

A review of the many scholarly attempts to determine the etiology of congenital dislocation of the hip leads to the conclusion there is not just one but multiple causes for this entity

The possibility of primary acetabular dysplasia has been supported by many It has been felt that the posterior superior buttress-like acetabular roof may not form on a genetic basis.

The femoral head originally must have been in relationship to the acetabulum since the joint develops as a cleft in the primordial cell block. Embryologic studies by Strayer reveal that this cleft does not become complete until the thirty five millimeter stage of the embryo is reached. Joint clefts have formed however even in the absence of the distal bone. It is obvious that, although the cleft may develop as the result of primary differentiating factors, the later development of the joint becomes more and more subject to other factors.

Badgeley reviewed the embryologic development of the hip joint beginning with the eight millimeter embryo The following comments are drawn from his article.

Only nerves and vessels are added from the trunk to the original limb bud in its development. At the fourteen and eight-tenths millimeters stage the predestined site of the hip joint can be seen as a dense accumulation of mesenchymal cells. These undifferentiated mesenchymal cells are still present at the twenty five millimeter stage between the femoral head and acetabulum. The capsular structures and glenoidal labrum have begun to form.

At the thirty three millimeter stage the ligamentum teres has appeared, and the joint space is completely differentiated. The glenoid labrum is prolonged over the head of the femur The limb bud has developed during this period in marked abduction. This is decreased considerably by the time the fifty-eight millimeter embryo is reached. During this developmental period the limb bud is also undergoing rotation.

Up to the point of innervation of the musculature of the limb it appears

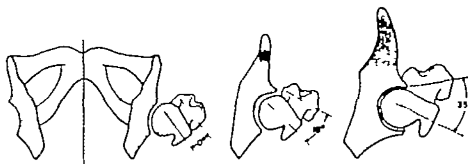


Figure 68 Development of the normal hip joint Note the progressive deepening of the acetabulum as the limb bud changes from a lateral to a parallel position There is progressive development of anteversion from 0 degrees *at three months to thirty five degrees* at birth (*Redrawn from* Badgley C E. Etiology of congenital dislocation of hip Bone & Joint Surg., 31A: 352 1949)

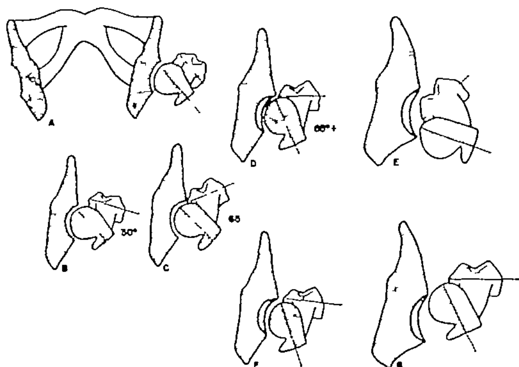


Figure 69 Development of congenital dislocation of the hip and acetabular dysplasia. A the thirty-five millimeter fetus without anteversion of the femur B and C development of normal and then excessive anteversion. D and E progressive dislocation with increasing anteversion, lack of normal pressure resulting in acetabular dysplasia F and G development of anterior dislocation. There is delay in proper timing of the rotation of the head into the acetabulum with both anterior and posterior dislocation as illustrated in the series from D to G so that at no time does it actually point into the acetabulum Dependent on the degree of the developmental error dysplasia or dislocation may be the result. (*Redrawn from* Badgley C E., J Bone & Joint Surg., Etiology of congenital dislocation of hip 31A: 353, 1949)

reasonable to assume that development of the joint is predestined. As Badgeley points out, delay in rotation of the limb bud or in innervation of the muscles at a period of rapid embryologic growth may produce alterations in the development of the acetabulum or in the head and neck of the femur Adaptive changes to malposition can occur in intrauterine as well as post natal life. Breech births have been noted frequently to be the subject of adduction contractures at the hip joint.

Underdevelopment of the limbus may be a factor in allowing dislocation of the hip The cartilaginous acetabulum and limbus cover at least half of the femoral head at birth Elongation of the capsule allowing subluxation and dislocation has been mentioned as a factor by Howorth

Anteversion, which may be the result of inward rotation of the limb bud as an angle of ninety degrees is traversed to bring the patella forward is a factor in poor presentation of the femoral head to the acetabulum. It may be one of the adaptive changes responsible at least in part for dislocation. The hip developing in the flexed position and extended after birth is aided in its tendency toward dislocation by the anteversion.

Dislocation of the hip does occur in association with other anomalies such as club feet club hands metatarsus varus and arthrogryposis. That dislocation may not be present at birth and yet develop presumably through the stages of dysplasia to subluxation to actual dislocation has been noted by many Such hips may, however already have joint development changes which will allow dislocation.

The development of dislocation after birth is similarly due to many factors. The classic direction of dislocation is said to be posterior although McCarroll states that primary anterior dislocation of the hip occurs in approximately one-fifth of the cases The presence of an adducted position of the femur relative to the pelvis with maintenance of this position by a myostatic contracture of the adductors stands out from a clinical point of view Enlargement of the ligamentum teres and of the Haversian pad of fat lying in the acetabulum depths has been implicated

The acetabulum appears to have the ability to respond to correct seating of the femoral head within it and is slow to develop when these forces are not transmitted to it. Anteversion of the femur as well as the adducted position are important in this regard

Weight bearing is regarded by some as a factor influencing a hip that is subluxated toward dislocation.

The forces of heredity may be instrumental in delaying rotation of the limb or its innervation and as such accomplish the sequence of events outlined by Badgeley However this is difficult to prove. Many factors appear to play a part in producing congenital dislocation of hte hip

The Acetabular Index

Severin has reported a means of measuring the acetabular angle originally devised by Hilgenreiner This is a generally accepted and simple measurement and serves to emphasize the importance of the acetabular obliquity in con



Figure 70 (left) Dislocated hip in newborn infant. Capsule opened from the anterior and superior aspects. The cartilage of the acetabulum instead of shelving over the femoral head is opposed to an enlarged ligamentum teres which intervenes between the cartilage and the femoral head Here the femur has been displaced from the acetabulum. *Figure 71 (right)* The normally developed hip—contralateral side of same patient. The head has been detached from the joint The cartilage of the acetabulum runs horizontally and shelves out over the femoral head



Figure 72 (left) The dislocated hip The enlarged ligamentum teres has been partially torn from its insertion The acetabulum runs vertically rather than horizontally An enlarged limbus was not seen The inability to seat the femoral head without division of the inferior capsule is quite evident. *Figure 73 (right)* The lower clamp is attached to the ligamentum teres which has been pulled inferiorly The joint space to accept the head is minimal, the acetabulum vertical, and the capsule tents across the joint just inferior to either end of the acetabular cartilage

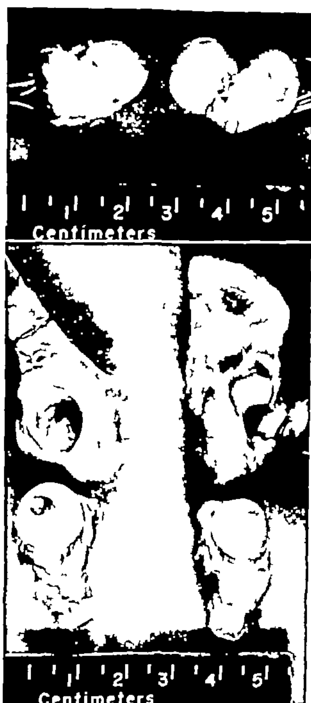


Figure 74 (top) View of superior aspect of femoral head, neck and greater trochanter showing deformity of head on left and increased anteversion. *Figure 75 (bottom)* Comparison of normal hip on left and dislocated hip on right with enlarged ligamentum teres on right pulled laterally

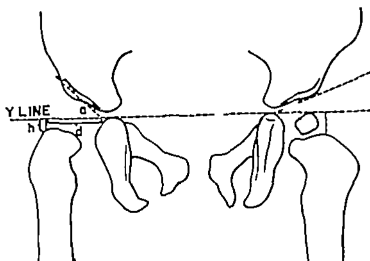


Figure 76 Measurement of the acetabular angle by the method of Hilgenreiner The y line connects the triradiate cartilages. Line h indicates the tendency of the diaphysis to ride superiorly as compared with the normal on the other side Line d indicates the lateral displacement form the acetabular floor (*Redrawn from* Hart, V L Congenital Dysplasia of the Hip Joint and Sequelae in the Newborn and Early Postnatal Life p 39 Charles C Thomas Springfield, Illinois, 1952)

genital dislocation of the hip Kleinberg and Lieberman report this measurement as twenty-seven and five-tenths degrees for the newborn and twenty degrees between age one and two The index is taken usually as normally ranging about twenty three degrees to twenty five degrees and over this point being abnormal particularly in the patient over six months of age. A line is drawn between the tips of each ilium at the tri radiate cartilage in the depth of the acetabulum. A second line drawn from the tip of the ilium to the outer edge of the acetabulum creates the angle known as the acetabular index.

The C E Angle

Wiberg has established an objective method of evaluating the position of the femoral head in relation to the acetabular roof The measurement is called the C E angle (center of the capital femoral epiphysis to the lateral edge of the acetabulum)

A line is drawn from the center of the head of the femur to the lateral edge of the acetabulum. This line forms an angle with a vertical line running through the center of the head A transparent circular protractor with its central point placed at the center of a circle enclosing the head of the femur readily enables this angle to be read when placed on the X ray film.

The angle will be less when either the acetabular roof is underdeveloped or when the femoral head is laterally displaced. It is helpful in relation to both acetabular dysplasia and subluxation of the femoral head.

Severin gives figures for this angle based on a study of one hundred and thirty six children between six and thirteen years. He felt that a value of less than fifteen degrees was definitely pathological. A C E angle of fifteen to nineteen degrees was equivocal and more than twenty degrees consistent with normal development.

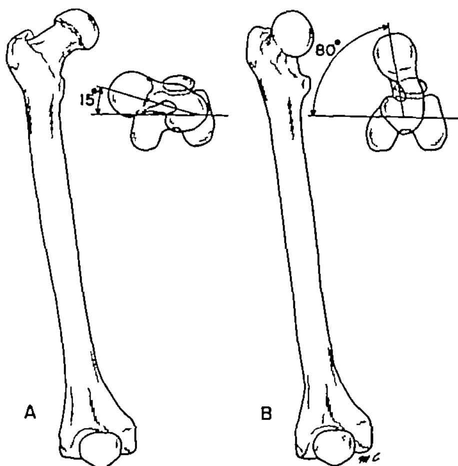


Figure 77 Anteversion of the femur illustrated diagrammatically as the relation between the axis of neck and the axis of the femoral condyles. In the antero-posterior view the femoral neck anteverted appears shortened and straightened in relation to the shaft.

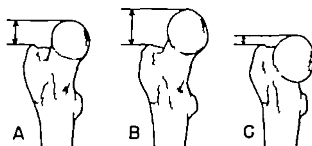


Figure 78 Anteversion (A) valgus (B) and varus (C) of the femoral neck can be differentiated in the antero-posterior view by the relative height of the femoral head above the greater trochanter

Anteversion

Anteversion may be defined as the relation of the long axis of the femoral neck to a line drawn through the femoral condyles in the coronal plane

Anteversion at birth is approximately twenty five degrees The tendency is for it to decrease with growth to five to fifteen degrees. Anteversion normally present is approximately fifteen degrees This is associated with about forty five degrees of internal rotation in flexion at the hip joint. When anteversion is increased internal rotation is correspondingly increased Thus excessive anteversion may permit internal rotation as great as ninety degrees.

Anteversion of the femur is recognized by the fact that a roentgenogram of the hip taken with the knees straight ahead reveals the hip to be in external rotation It is differentiated from valgus of the femoral neck by the fact that the height of the femoral head is not increased over the trochanter A roentgenogram taken with the femur in internal rotation is also helpful in differentiating from valgus.

Anteversion of the femur may play a part in poor development of the acetabulum after the femoral head has been placed in the acetabulum. In internal rotation the femoral head is well placed beneath the overhanging roof of the acetabulum. With the limb in the neutral position and the knee pointing forward anteversion of the femur will tend to bring the hip to the anterior lip of the acetabulum and tend to subluxate outward

Relation of the Hip to the Spine

Routine X rays of the spine taken of patients suspected of congenital dislocation of the hip is illuminating Two types of spine curvatures are often revealed

(1) *Curvature of the spine secondary to adduction contracture of the hip* The patient when aligned, in the anatomical position for the roentgenograms, will develop a pelvic tilt as the result of the adduction contracture. The pelvis will be "high on the adducted side This will result in a secondary curve in the low lumbar area to compensate for this tilt and align the occiput with the pelvis.

(2) *Curvature of the spine primary in the spine* This curve is a total curve involving the entire spine in a single long curve. This will result in a tilted pelvis and an adducted position of the hip Clinically, such babies will have a limitation of lateral bend in the direction correcting the curve There may or may not be an accompanying adduction contracture of the hip This primary spine curve is presumably the result of position in utero

When there is no adduction contracture, the hip may be quite well developed by X ray which accounts for the baby who is thought to have a congenital hip problem and yet little is revealed by roentgenogram. The thigh folds are often asymmetrical in such a child.

When there is an adduction contracture the hip is usually underdeveloped, i.e there is an increased slant to the acetabulum and delay in mineralization of the growth center of the proximal femoral epiphysis The improvement in such a hip will be markedly aided by trunk stretching exercises at the hip Failure to correct the trunk curve will result in delay in return of the hip to a normal appearance by X ray

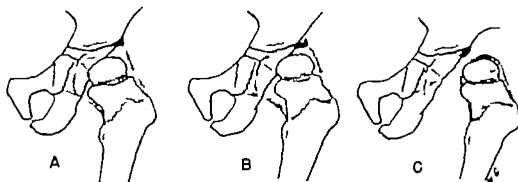


Figure 79 Dysplasia, subluxation and dislocation of the hip The limbus in subluxation is pushed superiorly against the ilium in dislocation the femoral head presses the limbus inferiorly into the acetabulum.

There are various forms of underdevelopment of the hip In the first year of life the hip may not develop into the stable functioning joint that it should be. This may be due to actual congenital deformity or neurologic conditions but much more commonly is due to a persistent adduction and anteversion of the hip The earlier that such abnormal stresses are recognized, the easier it is to correct the hip with further growth

The various stages of failure of the hip joint to develop may be divided into (1) The dysplastic hip (2) The subluxated hip (3) The dislocated hip The relationship between the underdeveloped hip joint and malum coxae senilis in later life has been pointed out by Vernon Hart. The severe disability that degenerative arthritis of the hip may impose on the adult makes the goal of normal hip development in the child a very worthwhile one to obtain.

THE DYSPLASTIC HIP

The mother frequently causes the inspection of these infants because of the difficulties noted with one leg or the other in placing diapers on the child. They are *recognized clinically by asymmetry of the thigh folds, an adduction con* tracture of the hip and apparent shortening of the leg. The infant is examined for an adduction contracture in recumbency In this position the thighs are flexed ninety degrees and then abducted. A limitation of full abduction is very evident by this examination.

A persistent adducted position of the femur relative to the pelvis may be due to a contracture of the adductor muscle group The adducted position may however be secondary to an elevation of one side of the pelvis with a curvature of the spine

The adducted position results in apparent shortening of the extremity The hip appears more prominent laterally on the involved side. The knee and popliteal creases are at different levels The inguinal fold is deepened on the involved side so that the labia is partially hidden by it.

Roentgen Signs

The acetabular roof has an angle greater than twenty five degrees The hip is not definitely displaced either laterally or superiorly There is usually recog



Figure 80 On attempted abduction in flexion an adduction contracture is demonstrated particularly on the right

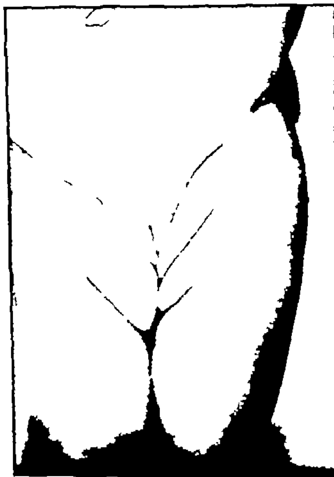


Figure 81 Inguinal fold obscuring labia on involved side Note asymmetrical folds.



Figure 89 Dysplasia of hip with underdevelopment of acetabulum

nizable anteversion of the femur A delay in development of the hip joint is evident. The ossification center for the femoral head is late in appearance Ossification of the acetabular roof is delayed along with delay in closure of the ischiopubic juncture.

Treatment

The treatment revolves around measures to eliminate the adducted position of the femur relative to the pelvis The inability of the trunk to bend equally to both sides must be corrected by stretching exercises if present

Tightness of the adductors is treated by stretching with the hips in flexion. In order to aid this stretching and maintain the abducted position a device to hold the legs apart is used. In very mild cases an extra diaper may be all that is necessary A greater measure of security in insuring correction is imparted by use of a Frejka pillow This device consists of a kapok pillow enclosed in a jumper The pillow is reasonably stiff waterproof and should equal in size the distance to the flexed knee on either side when the hips are abducted.

Such measures ordinarily result in the correction of the dysplasia of the joint in a three- to four month period when the abduction device is worn both night and day

THE SUBLUXATED HIP

Subluxation of the hip as a stage in the development of dislocation and as an entity unto itself has been studied by Hart. He notes that the untreated

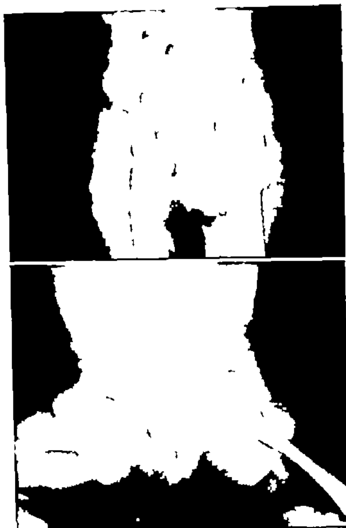


Figure 53 Subluxation of hip—note malalignment on abduction.

subluxated state may not go on to dislocation but remains with underdeveloped slanting acetabular roof and a tendency for femoral head to progress still further laterally and superiorly in the joint.

In this condition the acetabular roof again has a marked slant increased over the usual accepted angle of twenty five degrees The epiphyseal growth center is delayed in its appearance. This hip is distinguished from the dysplastic hip, however in that it rides laterally in the joint to the point of actual dislocation. These criteria form a triad enunciated by Putti to enable determination of the so-called "preluxation" state.

From the pathologic point of view the head of the femur may have pressed the cartilaginous rim or limbus of the acetabulum upward and toward the ilium. As brought out by Leveuf the limbus may remain attached in this position and be a serious detriment to the formation of the acetabular roof The acetabulum is shallow and the extent to which the acetabular roof overhangs the femoral head is minimal, the C E angle is decreased. The capsule is stretched but has not gone on to the hour-glass constriction or adherence of the capsule over the



Figure 84 Further development of congenital hip disease with riding upward of the femur

acetabulum except in its superior portion where it may be pressed between the femoral head and ilium The capsule pressed against the ilium may become converted to fibrocartilage.

The X rays mirror this change with marked widening of the joint. A line drawn through the center of the neck may no longer point toward the center of the tri radiate cartilage in the depths of the acetabulum. In other words there may be a subluxation proximalward resulting in the first stage of the so-called ' wandering acetabulum.

Clinical Signs

The adduction contracture or persistent adducted position of the femur relative to the pelvis is present. Asymmetrical thighs and buttock folds often accompany the condition. The trochanter is prominent on the involved side and aids in distinguishing the subluxated from the dysplastic hip Palpation of the femoral head beneath the femoral artery just below the inguinal ligament reveals that it is present in distinction to actual dislocation.

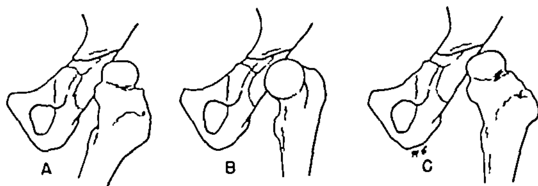


Figure 85 Some of the causes of poor seating of the femoral head in the acetabulum A adduction. B anteversion. C improper alignment.



Figure 86 Frejka Pillow consisting of stiff felt or kapok pillow enclosed in jumper

Treatment

If limitation of lateral bend is present in the trunk, it should be corrected by stretching exercises. The adduction contracture can be stretched gently by abducting the hip when it is in ninety degrees of flexion, and can usually be corrected fully in six weeks time An efficient device holding the thighs fully abducted is necessary for at least a three-month period—the exact time for discontinuance can be determined by the development of a normal joint by X ray Such a device may be a kapok pillow held between the legs in a jumper or straps Other methods include a plaster cast or an abduction frame. The hip should be fully corrected before the child is allowed to stand and start walking. Undercorrected subluxations do not regress but usually tend to progress after walking is allowed Anteversion, if a factor should be corrected by osteotomy

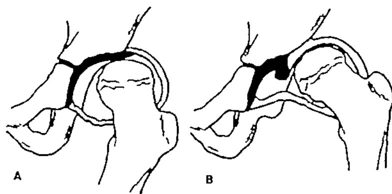


Figure 87 In A the femoral head is subluxated pressing the limbus superiorly In B the femur is dislocated pressing the acetabular rim inferiorly (*Redrawn from* Hass J Congenital Dislocation of the Hip p 48 Charles C Thomas, Springfield Illinois, 1951.

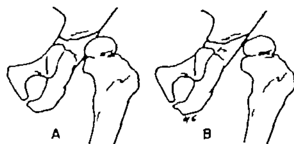


Figure 88 Undercorrected subluxation *tends to progress not regress.*

THE DISLOCATED HIP

The clinical signs that differentiate dislocation from the dysplastic and subluxated hip are those that follow the riding superiorly of the hip in relation to the acetabulum. These include shortening to measurement, telescoping, a positive Galeazzi sign failure to palpate the femoral head beneath the femoral artery in its normal anatomical position and the 'sign of the jerk.

Pathology

The hip, when dislocated lies superior to the acetabulum separating the gluteus medius and minimus from the ilium. The cartilaginous limbus forming the rim of the acetabulum lies inferior to the femoral head. Inversion of the limbus was noted in all cases in a series of twenty three open reductions by Somerville. The limbus is often indented by pressure from the head. It is normally largest posteriorly and superiorly and may be excessively large in dislocations The capsule is stretched to continue to enclose the head in its position distant from the acetabulum. There tends to be a constriction of the capsule between the head and the acetabulum. The capsule is often adherent to the ilium at least in part, where it is held against the bone by the pressure of the head. It may also be adherent inferiorly where it tents across the acetabulum.

Anteversion of the femur of some degree is a virtually constant accompaniment. Normal anteversion at birth is about twenty five degrees decreasing thereafter with growth to about fifteen degrees. The acetabulum may be filled



Figure 89 Child age 2 with bilateral dislocation of the hips, wide perineum and prominent trochanters.



Figure 90 In unilateral dislocation, flexion of the thighs to a right angle may reveal shortening on sighting along top of knees. (Galeazzi's sign)

by an enlarged Haversian fat pad. The ligamentum teres greatly elongated in actual dislocation may appear hypertrophied

In late dislocations the head may lose its round full shape becoming flattened on the area pressed against the ilium and assuming a more cone-like appearance The cartilage of the head may become pitted and fibrillated.

Clinical Picture

The patient is usually a female with inability to abduct the hip This is often noted when putting on the diaper by the mother Some asymmetry of the thigh and buttock folds may also be noted. Apparent shortening of the one leg is a frequent complaint.

The adductors are short in relation to the other musculature about the hip This is best demonstrated with the leg in ninety degrees of flexion at the hip when abduction it attempted. While forty five degrees or more of abduction is usually possible Infants with subluxed or dislocated hips have a limitation to twenty degrees or less.

The buttock fold on the side of the adducted hip tends to be higher with

asymmetry of the thigh folds below this point. The side involved has some apparent shortening and a high pelvis can be demonstrated on the involved side by palpating the anterior superior spines of the ilium. The inguinal fold runs into the labia making it appear smaller on the involved side.

With actual dislocation, there is absence of the femoral head from its position beneath the femoral artery just distal to the inguinal ligament. The trochanter is unduly prominent and lies above Nelaton's line connecting the anterior superior spine and the tuberosity of the ischium In unilateral hips there is a positive Galeazzi sign When the femurs are flexed to ninety degrees, one knee, due to the shortening of the thigh with dislocation, lies below the level of the other Internal rotation is usually increased to seventy degrees or more due to anteversion of the hip The "sign of the jerk" is the click that is felt when the thigh is abducted in flexion and *the femoral head slides over the* acetabular rim The adductors must be relaxed to elicit this click which may be felt on both exit and entry in the dysplastic acetabulum.

When bilateral, the perineum is widened It may be possible to detect telescoping of the hip by applying traction when the other hand grasps the pelvis with a finger palpating the trochanter

X rays reveal the hip to have slipped superiorly in relation to the acetabulum, *resulting in discontinuity in Shenton's line. Shenton's line is formed by the* inferior border of the neck and the superior border of the obturator foramen. *It is not a reliable sign of hip difficulty* when the only finding, but as an adjunct aids in the x ray interpretation. The dislocation may have begun anteriorly or posteriorly Acetabular views of the hip reveal the anterior or posterior relation to the acetabulum. The signs of underdevelopment of the joint and proximal femur previously discussed are also present.

In so-called unilateral dislocation the contralateral hip is worthy of study A roentgenographic study by DiPrampero has brought some interesting facts to light in this regard. Two hundred patients with unilateral dislocation were reviewed. Subluxation or dysplasia of the hip was found in one hundred and eight cases on the contralateral side.

Signs Due to Weakness of the Hip Abductors

The displacement upward of the femur and lack of a stable joint results in weakness of the hip abductors, of these the principal one is the gluteus medius. Shortening of the functional *length of the muscle results in weakness.*

The inability of the abductors to elevate the pelvis on the opposite side when standing on one leg results in a positive Trendelenberg sign. On standing on one leg the pelvis drops on the opposite side. This can be made more apparent by marking the posterior superior spines of the ilium before the test is performed.

This inability to elevate the pelvis results secondarily in inability to clear the opposite leg on walking. In order to clear the leg when walking the patient compensates for the abductor weakness by bringing the body weight over and beyond the femoral head. *This allows the lateral* trunk musculature to aid in elevating the pelvis

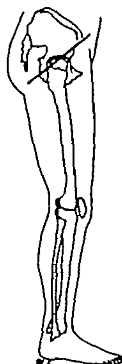


Figure 91 Diagram of Nelaton's line running from the anterior-superior spine of the ilium to the tuberosity of the ischium In dislocation the greater trochanter is palpated above this line

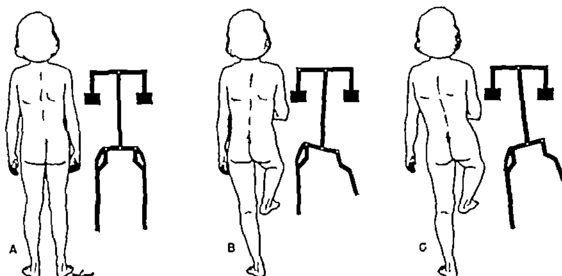


Figure 92 Signs of gluteus medius weakness such as may be associated with congenital dislocation of the hip A normal standing. B dropping of pelvis of contralateral side when standing on affected leg C compensation for weakness when walking by shifting center of gravity of trunk in order to elevate pelvis on opposite side

Such a gait transferring the trunk over the involved hip with each step results in a medius limp If both hips are involved the trunk is alternately transformed over first one hip and then the other The symmetry of such a gait makes it not as noticeable as in cases where only one hip is involved and accounts for the fact that children who are not found to have hip dislocation until they are walking are discovered earlier with a unilateral rather than a bilateral hip problem.

The Differential Diagnosis

Since the adducted position of the femur relative to the pelvis is responsible for most of the clinical signs that make us suspicious of dysplasia subluxation, or dislocation of the hip, a review of other conditions which might cause this state results in a surprising list. All of these conditions obviously have other means available in their examination which allow differentiation so they are presented as a list, which brings their occurrence to mind.

The attempt to abduct the hip either on flexion or extension may reveal a limitation in this plane. Such a limitation could be due to

(1) Voluntary resistance—The thoroughness and persistence of the examination will overcome this

(2) Spina bifida—with weak hip abductors and adduction contracture develops with persistent maintenance of the adducted position

(3) *Hip joint infection—pyogenic, tuberculosis, luetic etc. Also has limited* motion in other places

(4) Obstetrical trauma—contusion, fractures of the epiphyseal area and *dislocation, may result in resistance to abduction while tender*

(5) Chondrodystrophy—Morquio's Disease and Hurler's syndrome may have limited abduction but also have other obvious stigmata on inspection.

(6) *Scurvy and rickets may result in painful limbs and limited motion.* Scurvy occasionally is associated with the reverse of this—a flaccid pseudoparalysis.

(7) *Developmental coxa vara—with deformity of the femoral* neck, this condition apes congenital dislocation well on examination but such a deformity is unusual below the age of four The femoral head is still in the acetabulum, and there is no telescoping.

(8) Newborns—limitation of full extension at the hip and knee is present at birth and limitation of abduction is associated with it. The examiner should be able to abduct each hip twenty degrees however

(9) Cerebral Palsy with spastic paralysis characteristically has a stretch reflex in the adductors which limits abduction.

(10) Still's Disease—rheumatoid arthritis in children involving the hips limits motion in other places in addition to abduction.

(11) Poliomyelitis—an adduction contracture may develop although an abduction contracture is more usual

(12) Congenital scoliosis may result in an elevation of the pelvis on one side and an adducted position of the hip but is ordinarily not associated with an actual limitation of motion of the joint.

Treatment of Dislocated Hips

The age group when the patient is presented for treatment makes a great difference in the ultimate outlook and *severity of the measures for* tment. An awareness of the condition by t community resul arly presentation of the patient for tr is preventive the *highest sense of the word.*

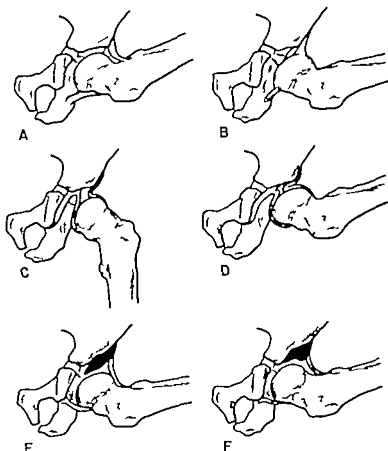


Figure 93 Possible obstacles to reduction include A and B adherent capsule C and D capsule lining the joint with hour glass constriction E and F inverted or enlarged limbus. (*Redrawn from* Hart, V L. Congenital Dysplasia of the Hip Joint and Sequelae in the Newborn and Early Postnatal Life p 48 Charles C Thomas, Springfield, Illinois, 1952.)

Dislocated Hips Below Age One

The object should be to procure an atraumatic reduction. In this age group it may be possible by elevating the trochanter gently as the thigh is flexed and then abducted to slide the femoral head into the acetabulum.

If this is not possible traction in line with the trunk in order to stretch the flexors may be necessary McCarroll recommends skeletal traction by means of a distal femoral pin placed proximal to the epiphyseal line in the age group over one. Once the femur appears to be pulled down in normal relationship to the acetabulum it is gradually abducted. Traction in abduction initially may not stretch the flexors and results in the femoral head being pressed tightly against the ilium or limbus

There are traction splints which are adjustable into abduction which work well but abduction without traction is not enough to secure a truly traumatic reduction

Once reduced the hip can be held in place by a bilateral short leg plaster spica holding the thighs in flexion and full abduction—after a period of several

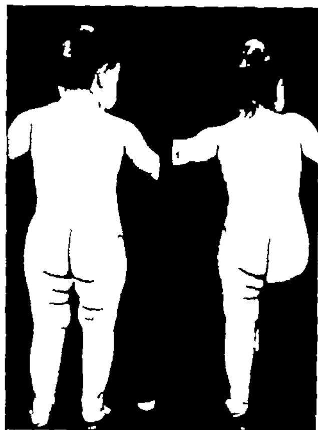


Figure 94 Dislocated hip on the left with high buttock fold, asymmetrical thigh folds, prominent trochanter and positive Trendelenberg with dropping of the opposite side of the pelvis when standing on the affected leg.

months an abduction splint such as that of Hass or Ilfeld may be all that is necessary to insure further development of the joint.

The roentgenograms of the hip should include antero-posterior and acetabular views and their review should be exceedingly critical. Failure to achieve a perfect reduction in both views should result in open reduction but this is seldom necessary in the infantile age group

Some infantile hips will readily reduce under anesthesia but forceful maneuvers should never be used to achieve this result. Gentle massage of the adductors may result in the ability to achieve full abduction.

Most patients with dislocated hips have excessive anteversion which prevents normal seating of the femoral head when the knee is foreward and the limb in line with the trunk. In the infants below six months there is no need to rush into an anteversion osteotomy since there is great opportunity for corrective growth in this rapidly growing period Once over this early period the need for osteotomy may be demonstrated by roentgenograms of the hip taken antero-posteriorly with the hip abducted and neutrally rotated and compared with films taken in internal rotation and abduction. Such films may reveal the femoral head to point out of the joint in neutral rotation and to be in perfect alignment in internal rotation Where necessary osteotomy should not be delayed.

Despite the apparent ease of treating early infantile hips, difficult cases are

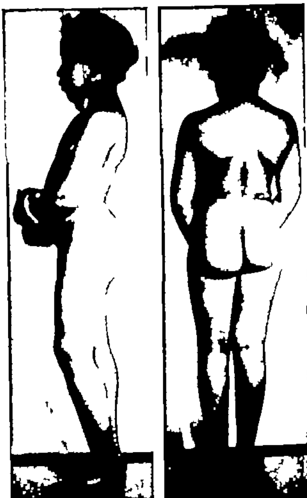


Figure 95 Congenital coxa vara present in this child mimics most of the physical signs of congenital dislocation of the hip with increased lumbar lordosis trochanters above Nelaton's line and gluteus medius weakness. The femoral head, however is palpated in normal position.

found, abduction is necessary for many months and careful roentgenographic examination must be made in order to procure a well developed joint with the femoral heads in normal position.

An additional aid in determining the adequacy of reduction in questionable hips consists of contrast injection. Approximately three cc s of an appropriate contrast medium to which the patient is not sensitive and which does not irritate the joint can be inserted into the joint cavity The approach can be lateral to the femoral artery and anteriorly or beneath the femoral artery through the hip adductors

The dye gives the most information if inserted just prior to reduction and films taken. The hip is then reduced. The position of the limbus can be determined as well as the presence of soft tissue filling the joint.

Dislocation of the Hip Over Age One

These children have been walking on the dislocated hip The result is considerable displacement compared to the previous age group

Traction preliminary to reduction should be routinely used to bring the hip opposite the acetabulum unless the mobility of the hip allows this when first seen Merely placing the hip in wide abduction fails to relieve the adduction contracture since the femoral head often slides further posteriorly and proximally

A more efficient form of traction consists of the use of a Thomas splint in mild abduction. The leg is literally drawn through the splint which is attached to adequate counter traction. Skeletal traction with femoral pin is usually necessary

Traction should be continued sufficiently long to allow an easy closed reduction The percentage of hips considered for open reduction can be expected to rise

After closed reduction the hip is placed in a plaster hip spica, holding the involved hip to the foot the uninvolved hip to the knee At the end of six weeks, the long leg is shortened to the knee so that rotation of the hip is possible. Such spicas usually last two to three months before the child outgrows them. When the spicas are discontinued is a matter of judgement based on a study of the X rays as the hip progresses The acetabular roof should develop into the normal range, and the epiphyseal growth center appear The ability of the acetabular roof to respond to the femoral head placed in normal position within it usually continues at least in part to age four

While in the cast the patient is kept on a Bradford Frame which eases the care of the child and keeps the cast clean. The frame has an opening beneath the genitalia and is kept at a slight downward slant so that urine will not run into the cast but into the bedpan beneath the frame.

Closed reduction should only be done gently when it is done. Bost *et al.* give the incidence of aseptic necrosis of the femoral head following closed reduction as twelve per cent in those cases done in the first year of life fifty nine per cent in the second year and sixty two per cent in the third year Aseptic necrosis following hip reduction does not always run the protracted course sometimes associated with Legg Perthes disease

Dislocated Hips Over Age Two

In this group skeletal traction and open reduction of the hip becomes the treatment for the majority of patients rather than the minority These youngsters are often bilateral cases since unilateral hips present more obvious physical signs in their gait and are therefore recognized earlier These children have been walking for a considerable period and considerable upward displacement of the hip is likely Telescoping of the hip is more easily elicited as a physical sign in this group than in any other

It becomes more important than ever to pull the femoral head into a position opposite the acetabulum. From this position regardless of the mode of therapy it can be placed in the acetabulum with relative ease if there is no obstruction. The efficiency of the traction becomes very important. Colonna has emphasized traction with the leg placed in a Thomas ring splint. The splint is a means of producing countertraction as the traction is applied to the leg.

There are some hips that can be replaced by closed reduction with no persistent widening of the joint and perfect alignment with the center of the head and neck pointed directly toward the triradiate cartilage in this age group

The majority of youngsters over two have developed changes in the capsule in the shape of the head and in adherance of the capsule to the acetabulum

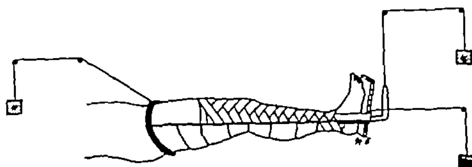


Figure 90 Traction applied to the hip to bring it down opposite the acetabulum Efficient counter traction is necessary to achieve results. Skeletal traction on the femur may be used

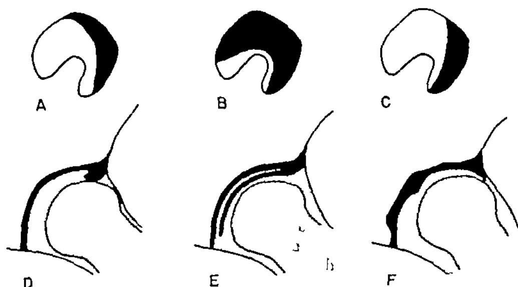


Figure 97 Possible situations of the cartilaginous limbus of the acetabulum which inverted may be an obstacle to reduction A B C Coverage of the acetabulum by the limbus which is most marked posteriorly D E, F Appearance of these situations in the arthrograms (*Redrawn from* Somerville E W Open Reduction in Congenital Dislocation of the Hip J Bone & Joint Surg 35B: 364 1953)

such that a perfect reduction is not obtainable Probably the most important barrier to a reduction is an enlarged limbus Open reduction is necessary to correct these difficulties.

An anterior ilio-femoral or Smith Peterson incision is often used The cartilaginous apophysis of the ilium is carefully spared as the gluteus group is removed from the ilium by subperiosteal dissection. The femoral head on opening of the capsule is found to have varying degrees of fibrillation of the cartilage and flattening of the surface opposed to the ilium The capsule is adherent to the side of the ilium and needs dissection to bring it down to the level of the actual acetabular rim The psoas muscle may run across the joint in a manner

preventing reduction and may need lengthening to remove it as an obstacle. The limbus of the acetabulum is visualized. It is frequently found to be large and inverted. Somerville found it a major cause of obstruction to reduction in each of twenty three cases of open reduction The limbus is largest posteriorly and extends anteriorly a variable distance. Removal of the excessive portion and fibro-adipose tissue in the joint results in a reduction being readily obtained. The hip usually appears most stable in internal rotation.

The femoral head, when placed in the acetabulum, is held in the position which is most stable. The capsule is repaired in this position.

A spica is applied, which is bivalved at three weeks post- operative, and gentle active guided exercises into further internal rotation and abduction begun. If the hip is placed in a position of extreme internal rotation the capsule tends to be wound about the neck constricting the circulation to the femoral head Such a position may be responsible for aseptic necrosis of the femoral head following open reduction.

Anteversion Osteotomy

Osteotomies to correct anteversion are only done in the presence of ex cessive internal rotation at the hip When done in the subtrochanteric region a Steinman pin is first placed in the femoral neck holding the hip in internal rotation. The distal fragment is then rotated until the knee joint is in line with the anatomical position of the body When the patient stands with the knee joint pointing straight ahead, the hip will then be in internal rotation by com parison with its previous position. Excessive internal rotation is no longer pos sible. The use of osteotomy to correct anteversion is a matter of judgement in each individual case since its degree will vary in each. Osteotomy can also be performed in the supracondylar region.

Hips that tend to redislocate after open reduction need correction of the factor allowing the redislocation. An acetabular shelf or buttress may have to be built and anteversion, if a factor corrected at a later date.

Treatment of Primary Anterior Congenital Dislocation

McCarroll has emphasized primary anterior dislocation of the hip as constituting twenty two and five tenths per cent of a series of congenital disloca tion of the hip Such dislocations may be the result of rudimentary development of the anterior portion of the acetabulum or the tendency of the acetabulum to face foreward thereby diminishing the anterior buttress in relation to the head. Dega in a review of one hundred fetal specimens noted that the angle of foreward inclination of the acetabulum was twenty nine and five tenths degrees.

McCarroll advocates preliminary skeletal traction for this condition followed by attempted closed reduction. Should the closed reduction be unsuccessful, open reduction is in order While an effort has been made to form either a shelf or buttress at the anterior margin, the results have not been entirely satisfactory If it is impossible to maintain excellence of reduction after an open procedure, it may be necessary to form a capsular arthroplasty type of procedure when the child is in the appropriate age group

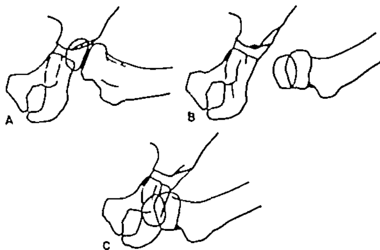


Figure 98 Incorrect positions following reduction. A femoral head posterior to the acetabulum B too great lataeral displacement remaining C inferior to the acetabulum in the region of the obturator foramen. (*Re drown* from Ham, J Congenital Dislocation of the Hip p 167 Charles C Thomas, Springfield, Illinois, 1951)

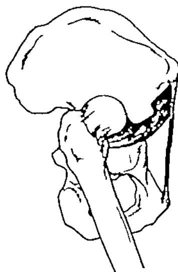


Figure 99 Constriction of the hip joint capsule by the tendon of the ilio-psoas in dislocated hip riding superiorly (*Redrawn from* Ham, J Congenital Dislocation of the Hip p 70 Charles C Thomas, Springfield Illinois 1951)

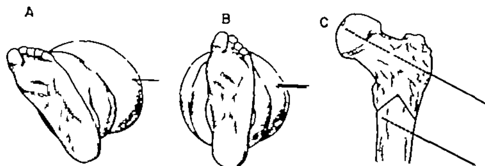


Figure 100 Anteversion osteotomy A the leg is internally rotated for desired degree of correction and Steinman pin placed in proximal fragment. B the bone is divided and distal fragment rotated until knee is upright—second pin is inserted in distal fragment parallel to first. C anterior view of final situation with parallel pins in place These are incorporated in plaster

Dislocation of the Hip Over the Age of Four

Dislocated hips in this age group have a poor outlook so far as development of the hip is concerned after a mere replacement in the acetabulum, even when done openly. The joint must be maintained in abduction many months, frequently with little or no improvement in the acetabulum itself.

Two procedures are available to develop the joint to the point where the hip will be maintained in position. Colonna has developed an operation for the dislocated hip in this age group of four to eight years. After preliminary traction so that the hip is opposite the acetabulum, an open reduction is performed. In this procedure the joint capsule is carefully dissected free of its adherence to the ilium and surrounding structures. The acetabulum is curetted so that a deep and adequate cavity is produced. The acetabular cartilage will be removed in this process, and the cartilage line at the junction of pubis, ischium, and ilium visualized.

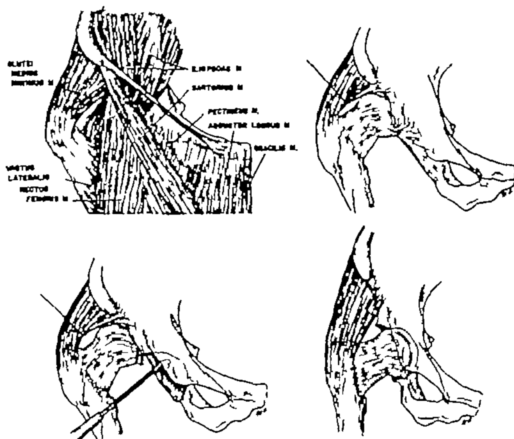


Figure 101. *Upper left:* in a capsular arthroplasty developed by Colonna, the intact capsule is exposed by a Smith-Peterson incision. *Upper right and lower left:* the acetabulum is refashioned by curetting and the femoral head enclosed in capsule is placed within the new joint. By preliminary traction the femoral head was brought down opposite the acetabulum so that it may readily be reduced. *Lower right:* the femoral head enclosed in capsular sac in place in newly fashioned acetabulum. (*Redrawn from* Colonna, P. C.: Congenital dislocation of the hip. American Academy of Orthopedic Surgeons Instructional Course Lectures, 8: 175, 1951.)

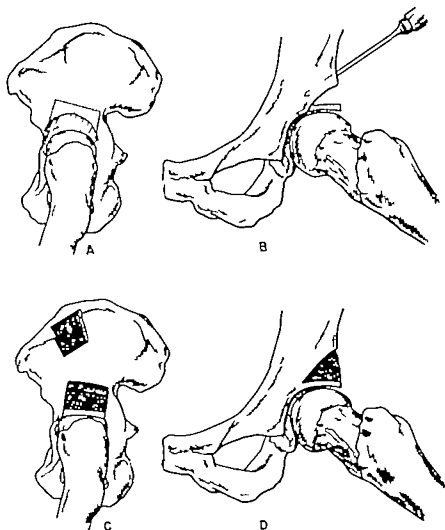


Figure 103 There are many methods of performing a shelf procedure to stabilize the femoral head in the true acetabulum. The operation basically pries down ilium and outer portion of the acetabulum and maintains the shelf in position by a bone graft which may be taken from the ilium (*Redrawn from* Hass, J Congenital Dislocation of the Hip p 288 Charles C Thomas, Springfield, Illinois, 1951)

The capsule which was left attached at the base of the neck is seen at its open end so as to enclose the femoral head The head and enclosing capsule is then replaced in the joint.

The patient is kept in a cast for four weeks thereafter Following this motion is begun at the hip in flexion abduction, and internal rotation with traction. There follows a very gradual and carefully guarded return to full weight bearing.

If traction preliminary to operation has brought the head opposite the acetabulum so that it is not forced into the newly made joint, the procedure is usually followed by a good result

A second procedure to create an adequate joint consists of building an acetabular shelf This procedure is of value also in treatment of the subluxated hip which is progressing to further subluxation. The insertion of a shelf may be enough to produce stability and prevent progression Again to insure a good result the hip must be pulled down by traction.

At operation the femoral head should lie opposite the acetabulum In Ghormley's procedure the anterior iliac crest and acetabular margins are exposed by a lateral hip incision The capsule is kept intact and bone chips are turned down over the head from the ilium leaving a bony notch The anterior superior spine and anterior portion of the ilium are freed subperiosteally This section of the ilium is then osteotomized on a level The leveled edge of this bone graft is then placed into the supra acetabular groove Traction is maintained for eight weeks with the patient in a spica to the ankle Crutch walking is then permitted. Where the procedure is being done for a hip drawn down opposite the acetabulum, a pitfall may be the failure to recognize the true acetabular rim at operation Not infrequently the capsule is turned up on the ilium and adherent to it. The capsule will frequently have to be dissected from the ilium until the true rim is reached. Occasionally as suggested by Anderson and Bickel, the capsule may be opened and the acetabular margin visualized

Variations of the method of building an acetabular shelf have been described by Alber, Gill Compere Phemister and Lowman Both the tibial grafts and sections of the ilium may be used as grafts after the superior posterior or anterior portions of the acetabulum are pried down over the femoral head by osteotome

There remains a small group of patients becoming quite rare in recent times who are in the older age group with a severely dislocated hip well removed from the original acetabulum. These hips are those that cannot be pulled down by traction to a point opposite the acetabulum. Here the general aim is to secure greater stability for the hip thus aiding the elimination of the medius limp

Exercises to develop greater strength in the gluteus medius are used. When bilateral these cases are perhaps best left alone. When unilateral, some stabilizing procedure is often used This includes the building of a shelf with the hip brought to a position just above the acetabulum after the suggestion of Dickson. Osteotomies have been devised by Lorenz and Schanz which procure increased bony support by displacement of the proximal portion of the distal fragment medially In the case of Lorenz osteotomy there is an actual placing of the shaft in the acetabulum.

Criteria for Reduction

Clinical

(1) Diminished prominence of the trochanter
(2) Palpation of the femoral head beneath the femoral artery one finger's breadth below the inguinal ligament
(3) Inability to extend the previously extending knee.

X Ray

(1) No widening of the joint space
(2) Line through the center of the femoral neck and head runs through the center of the tri radiate cartilage in all views.

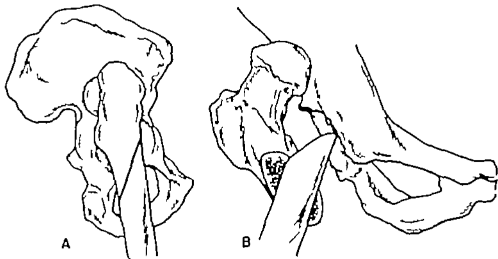


Figure 103 Osteotomy may be used to increase stability in an old still dislocated hip In the Lorenz osteotomy the shaft of the femur is displaced into the acetabulum. (*Redrawn from* Hass, J Congenital Dislocation of the Hip p 205 Charles C Thomas, Springfield Illinois, 1951)

NEUROGENIC DISLOCATION

Such hip dislocations occur following both spastic and flail paralysis of the lower extremities. The underlying etiologic factor is often a combination of several events associated with each form of difficulty

A principal factor in association with spastic paralysis is the persistent adducted position. Adductor shortening with use, and the maintenance of this position with growth may result in gradual superior bending of the limbus and riding out of the femoral head. This is aided by valgus of the femoral neck and uncorrected anteversion.

In flail paralysis with non weight bearing valgus of the femoral neck is a very common finding. Uncorrected anteversion of the femur may accompany it. One is hard put to properly weigh valgus and anteversion as to their relative importance in any one case. In poliomyelitis where the muscles may not be completely flail and contractures arise, these may also play a part. The importance of the tight fascia lata with contracture in abduction results in a flexed position with the hip in neutral. Extension then tends to drive the hip posteriorly and over the acetabular rim. Poor seating of the femoral head due to valgus or anteversion aids the progress of the hip laterally Again, however, factors such as anteversion and valgus usually must exist in combination to result in actual dislocation.

Neurogenic dislocations are seen particularly in connection with myelomeningocele, cerebral palsy or poliomyelitis.

Treatment

The maintenance of an abducted position helps to counteract the progression of the femoral head toward dislocation It also aids in maintaining reduction once this has been achieved. The forces which originally drove the femur into



Figure 104 Neurogenic dislocation on right with excessive anteversion and valgus in both hips



Figure 105 Anteversion osteotomy on right to aid in keeping femoral head in normal relation to acetabulum

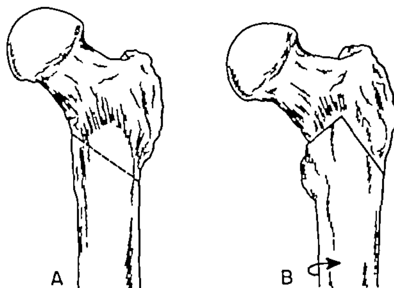


Figure 100 A type of osteotomy for anteversion which also corrects valgus and is of special help in neurogenic dislocation.

a dislocated position must be corrected however, to guard against recurrence. Keeping the patient in plaster spicas over the years may be too costly from the point of view of mental and physical health and other means must be sought.

In the presence of a well developed acetabulum correction of the anteversion and valgus by femoral osteotomy may be all that is necessary In cases where the acetabulum is deficient shelf procedures to create a stable abuttment may be indicated. Levering down the acetabular rim and blocking it in position will only be successful if the hip has been pulled down by traction so that it is not exerting a strong pressure on the reconstructed area.

References

ADAMS F The Genuine Works of Hippocrates. London, 1849

BADGELEY C E. Correlation of clinical and anatomical parts leading to a conception of the etiology of congenital hip dislocation. J Bone & Joint Surg., 31 A: 341 1949

BADGELEY C E. Etiology of congenital dislocation of the hip J Bone & Joint Surg., 31-A: 341 1949

BOST F C HAGEY H SCHOTTSTAEDT E R., AND LARSEN L. J The results of treatment of congenital dislocation of the hip in infancy J Bone & Joint Surg., 30-A: 454, 1948

BRADFORD E H Treatment of congenital dislocation of the hip Boston Med. & Surg. Jour., 108: 73 1883

BRADFORD E. H Congenital dislocation of the hip—results of treatment at the Boston Children's Hospital Am. Jour Orthop Surg., 7: 57 1909

COLONNA P C An arthroplastic operation for congenital dislocation of the hip—a two stage procedure Surg. Gynec. & Obstet., 63: 777 1936

COLONNA, P C Arthroplasty of the hip for congenital dislocation in children J Bone & Joint Surg 29: 711 1947

COMPERE, E. L. The shelf operation for congenital dislocation of the hip Am J Surg., 43: 401 1939

CREGO C H Jr Preliminary skeletal traction in the treatment of congenital dislocation of the hip South M J., 26: 845 1933

CREGO C H Jr., AND SCHWARTZMANN J R Follow-up study of the early treatment of

congenital dislocation of the hip J Bone & Joint Surg., 30-A: 128, 1948.
DeNucé M Luxation congenitale de la hanche Operation de Hoffa. Rev d'orthop 1a.4: 108 1893
Dickson F D The shelf operation in the treatment of congenital dislocation of the hip J Bone & Joint Surg., 17: 43 1935
DiPrampero A. Morfolojia radioghica dell'ance con detta sana nella lussazione congenita laterale Chir d'org di Movimento, 25: 1 1930
Frejka, B Prävention der angeborenen Juftöerrenburg durch das Abduktionspolster Wien. klin. Wchnschr., 91: 523 1941
Galeazzi, R. Eiuti lontani della cruta incrimenta della lussazione congenita dell'anca. Arch Ortop., 32: 191 1915
Ghormley R. K. Use of the anterior superior spine and crest of the ilium in surgery of the hip joint. J Bone & Joint Surg., 13: 781 1931
Gill, A B Plastic construction of an acetabulum in congenital dislocation of the hip. J Bone & Joint Surg., 17: 48, 1935
Gill, A. B The end results of early treatment of congenital dislocation of the hip. J Bone & Joint Surg., 30-A: 442 1948
Hart V L. Primary genetic dysplasia of the hip with or without classical dislocation. J Bone & Joint Surg 24: 753 1942.
Hart V L. Congenital dysplasia of the hip joint and sequelae Charles C Thomas, 1952.
Leveuf J Primary congenital subluxation of the hip J Bone & Joint Surg 30-A: 875 1948.
Lorenz, A. La riduzione della lussazione congenita dell'anca. Arch. di Ortop 14: 1, 1897
McCarroll, H R. Primary anterior congenital dislocation of the hip J Bone & Joint Surg 416 1948
Muller, G M and Seddon H. J Late results of treatment of congenital subluxation of the hip J Bone & Joint Surg 35-B: 342, 1953
Nelaton E. Elements de pathologie chirurgicale edit. Paris Pean, 3: 323, 1874.
Poli, A Contributo alla cura precoce della lussazione congenita dell'anca. Arch. Ortop 52: 749 1936.
Putti V Early treatment of congenital dislocation of the hip J Bone & Joint Surg., 11: 798 1929
Severin E. Arthrography in congenital dislocation of the hip J Bone & Joint Surg., 21: 304 1930
Severin E. Contribution to knowledge of congenital dislocation of the hip joint. Late results of closed reduction. Acta Chir Scandinavia 84: Supplementum 63, 1941
Smith Petersen W N A new supra-articular subperiosteal approach to the hip joint. Am. J Orthop Surg 15: 592 1917
Somerville, E. W Development of congenital dislocation of the hip J Bone & Joint Surg., 35-B: 368 1953
Somerville, E. W Open reduction in congenital dislocation of the hip J Bone & Joint Surg 35-B: 363, 1953
Wiberg G Studies on dysplastic acetabular and congenital subluxation of the hip joint. With special reference to the complications of osteoarthritis. Acta Chir Scandinav 83: Suppl. 58, 7 1939

DEVELOPMENTAL COXA VARA

Developmental coxa vara has been related to congenital short femur by Golding. It is not unusual to find the two co-existing. Congenital coxa vara results from anomalous ossification of the proximal femur The deformity thus produced gives rise to a medius type limp and marked limitation of abduction. This limitation, when bilateral is especially disabling.

The lesion results in a progressive deformity Abnormality of the gait does not ordinarily become noticeable before the age of three or four Thereafter,

the gait difficulty caused by the mechanical abnormality of the proximal femur is increasingly noticeable. The disease is equally divided in so far as unilaterality, bilaterality, and sex are concerned

Roentgen Findings

The usual angle formed by the femoral neck with the shaft of the femur is 135 Some change in the direction of increasing varus is always present, and if the patient is first seen in the four to seven age group, the neck usually forms a right angle with the shaft. The area occupied by the epiphyseal line is unusually wide in the early age group This increased area of radio-lucency apparently results from the epiphyseal line plus a gap just distal to it, which occasionally branches away from it as it nears the base of the neck. A triangular segment of bone is thus isolated as emphasized by Le Mesurier

In later cases the area occupied by the epiphyseal line becomes quite thin, irregular and fragmented. The neck appears shorter than would normally be expected. The line tends to close prematurely It becomes evident that there is a decreased rate of growth at this line.

The trochanteric epiphyseal line appears to be normal.

Clinical Picture

The child is brought to medical attention because of the waddling gait due to the medius limp The limp is progressively more noticeable as the age advances Pain is not a symptom.

The limitation of abduction is the most striking of the affects on the hip motion, itself since the other planes of motion are usually less affected, except with severe deformity There is a positive Trendelenberg on standing on the involved side. The trunk shifts over the involved hip with weight bearing in order to clear the opposite leg and elevate the pelvis, giving rise to the gait associated with weakness of the gluteus medius The trochanter is above Nelaton s line. The condition may be confused with dislocation of the hip on physical examination Actual dislocation is a much more common lesion. The femoral head is palpated in normal position Telescoping cannot be elicited in coxa vara however Shortening to measurement would depend on the degree of coxa vara present.

Course

If the case is untreated, progressive deformity can be expected to at least a right angle, and more often an even more severe change in femoral neck shaft relationship in the direction of varus.

Since premature closure of the epiphyseal line is the rule it is unusual to see progression after the age of twelve. Once the line is closed the neck is composed of bone of apparently normal tissue

Pathology

There has not been a good opportunity to study the actual pathology involving the femoral neck. Study of the x ray picture yields the principal in



Figure 107 Bilateral developmental coxa vara Note widened epiphyseal line and filling in of inferior neck angle with triangular area of ossification.

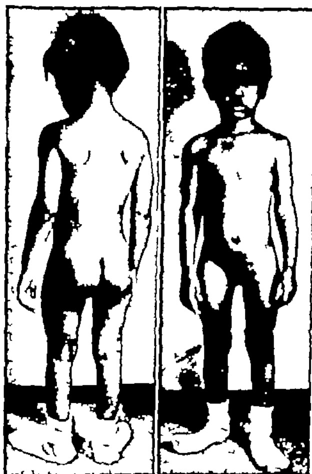


Figure 108 Child with bilateral developmental coxa vara and clinical signs of widened perineum and prominent trochanters.



Figure 109 Progression in developmental coxa vara There is an eight year interval between the roentgenogram on the left and that of the same hip on the right with marked increase in deformity

formation about the disease It is presumed to be a form anomalous ossification of the proximal third of the femur

Treatment

Traction has not been shown to affect the deformity The treatment would depend on the deformity at the time the patient is first seen the amount of growth left and the gap present in the femoral neck.

Early Cases

Where the deformity present is compatible with good function of the hip and further progression can be expected, attention may be directed to attempting to gain rigid fusion of the epiphyseal line Le Mesurier has advocated both nailing with a Smith Peterson nail inserted across this gap and actual bone grafting. Bone grafts may be inserted from the lateral cortex and passed up the femoral neck. Such cases should be followed through the growth period thereafter since continued growth of the trochanteric epiphysis may still result in a sufficient varus deformity to need osteotomy

Late Cases

An abduction osteotomy which obtains adequate correction does not give an absolutely perfect hip so far as motion is concerned but results in im

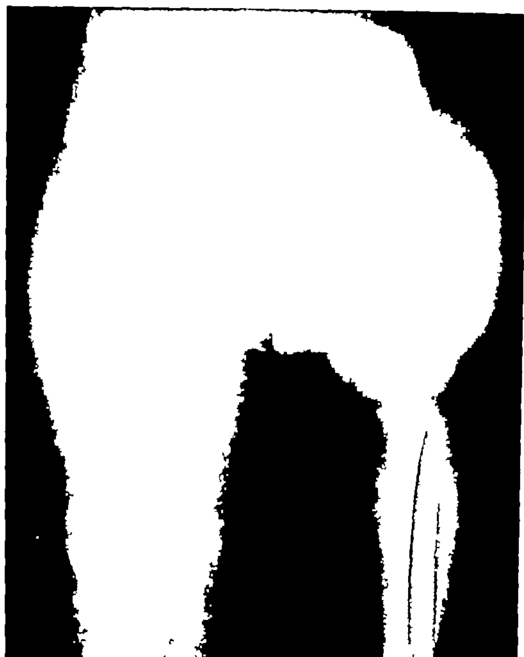


Figure 110 Congenital short femur and developmental coxa vara in combination

provement in gait and abduction at the hip Where the condition is bilateral such improvement may be essential. The osteotomy is done just below the level of the lesser trochanter through a lateral incision. The proximal fragment is fixed with a Steinman pin running up the femoral neck and across the epiphyseal line The fragments are kept end to end and not allowed to override. The amount of abduction necessary for correction should be previously measured on the x ray film.

Because of the lengthening of the leg which results with abduction, it is

difficult to obtain and maintain the desired correction Traction may be used as an aid, but the proximal fragment must be controlled with external fixation

The spica cast may be wedged bringing the distal fragment into further corrective abduction post operatively Again the proximal fragment must be controlled.

Good healing is obtained before the cast is removed and the hip is then mobilized with the aid of traction and exercises

References

BARR, J S Congenital coxa vara, Arch. of Surg., 18: 1909 1929

GOLDING F E. Congenital coxa vara J Bone & Joint Surg 30-B: 161 1948

LE MESURIER, A. B Developmental coxa vara, J Bone & Joint Surg., 30-B: 595 1948

SLIPPED CAPITAL FEMORAL EPIPHYSIS

The essence of slipped capital femoral epiphysis consists of a mechanical change from the normal of the relationship of the femoral neck to the capital femoral epiphysis. It occurs primarily in adolescents

The results of treatment in cases where a minimal change has taken place are better than those following a marked change in the relationship This places great value on early diagnosis.

The usual age group is ten to seventeen years, with females by virtue of the approximate two year advancement in skeletal maturation being seen in the early portion of this group Rarely even younger cases are seen. It is much more common in males (five to one) and bilateral cases occur in approximately twenty per cent. Klein *et al* state that x ray signs of bilateral involvement exist in forty per cent, even though no clinical symptoms may exist.

Etiology

Two general body types are frequently involved by this disease One is the so-called Froehlich type with female fat distribution and underdevelopment of the genitalia the other often seen is the thin individual with long extremities and very rapid adolescent growth Approximately one-half the patients seen with this disease fall into one or the other group

The epiphyseal line in the femoral neck is perhaps more subject to a shearing type of stress than any other and while it is possible to visualize the onset and progress of the disease on the basis of these factors alone, this is as yet unproved

An endocrine basis for this disease is suggested by the growth abnormalities Both growth hormone and sex hormone affect the rate of proliferation of cartilage cells at the epiphyseal plates The rate of proliferation of these cells may exert a profound affect on the rate of skeletal growth.

Harris tested the shearing force necessary to detach the proximal tibial epiphysis from the shaft in rats. In growth hormone treated rats with epiphyseal plates thickened principally in the layers of proliferating and maturing cartilage cells a lesser than normal average force was required In estrogen treated

rats with thinner plates at the epiphyseal line a greater average force was necessary than that used in the normal controls.

While all epiphyses presumably react to endocrine changes, the placement of the proximal femoral epiphysis and consequent subjection to shearing stress may result in this area becoming involved to the exclusion of others.

Ghormley brought out the fact that the epiphyseal plate changes position from horizontal to oblique during pre-adolescent and adolescent periods.

There are some who favour trauma taking place at this point of low resistance to shearing stress as the only etiology in view of a lack of clear-cut evidence for endocrine factors

Pathology

Irritation of the hip joint results in edema and hyperemia in the synovia. In the earliest stage of slipped epiphysis these changes plus some perivascular lymphocytic infiltrations and villous formation are all that are noted In the cases of actual slipping the epiphysis is not completely detached from the neck but remains attached by periosteum and fibrous tissue. The epiphyseal disc remains attached to the head, the slipping taking place in the more distal layers The epiphysis is downward and posterior to the neck which slips forward and upward The angles between neck and epiphysis tend to fill in with callus. The articular cartilage appears normal. The bluish fibrous tissue covering that part of the neck formerly contiguous with the epiphyseal line is smooth glistening and tends to hide the true location of the epiphyseal line. Over a period of several years the head and neck go on to bony union with incorporation of the epiphyseal line. Late changes in this disease reveal a firm, sclerotic and thickened capsule and conversion of the callus at the inferior angle of the epiphysis to bone

Clinical Picture

The typical patient has noted pain in hip thigh or knee often for several months and developed an antalgic limp Since the hip capsule may not always be markedly irritated it is not uncommon for the patient to have a history of low grade symptoms of considerable duration.

If the symptoms and gait are acute one suspects slip with the neck and shaft rotating outward and upward in relation to the head.

On examination the appearance of the patient may be so typical as to allow suspicion of the diagnosis on sight. The large frame, heavy thighs underdeveloped genitalia and leg lying in external rotation point to the fact of slipped epiphysis When the thigh is flexed the leg tends to ride into external rotation and frequently the leg cannot be brought to neutral.

If the hip is irritated other motion will be limited as well and a hip flexion contracture present. If hip irritation is minimal the patient may actually have increased hyperextension consistent with his degree of slip

Suspicion of slipped femoral epiphysis should result in the patient leaving the examining room in recumbency since further slip complicates treatment and jeopardizes the end result.

Figure 111 A well defined epiphyseal slip is seen on the right with widened epiphyseal line and a build-up of bone beneath the head on the inferior neck.



Figure 112 Acute slip of the femoral epiphysis without ossification at the inferior neck

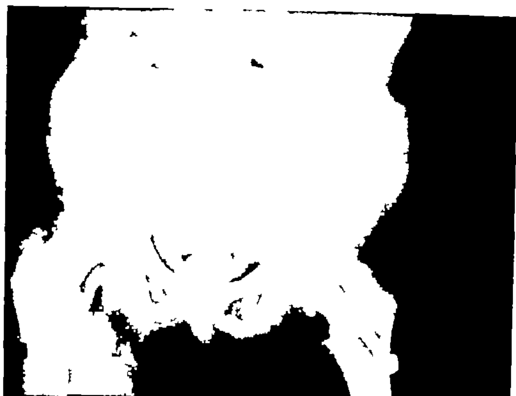




Figure 113 Upper slipped femoral epiphysis treated by screw fixation on the right with apparently normal hip on left. *Lower* same patient four months later with filling in of the epiphyseal line on the right, but developing widening and radiolucency of the epiphyseal line on the left.

Roentgen Signs

The earliest manifestation of slipping consists of widening of the epiphyseal line and irregular demineralization of the area immediately adjacent to it The capsular shadow is rounded and more prominent than normal

Klein *et al* have emphasized the fact that a line drawn along the superior surface of the neck to the outer edge of the acetabulum normally leaves the lateral angle of the head outside it Medial slipping can be detected in the anterior posterior view of the hip when the head is not transected by this line but lies within it. Posterior slipping is detected in the lateral view when the change in relationship is more obvious as the axis of the neck leaves the epiphysis A generalized atrophy or demineralization of the head and neck may be apparent.

The epiphysis does not always lie both posterior and medially in relation to the neck In the AP view in Klein's case a medial slip was apparent in only sixty eight per cent. In the lateral view of these cases posterior displacement was evident in ninety-eight per cent In speaking of the degree or distance of slip the lateral view is ordinarily used.

Treatment

Treatment begins immediately upon suspicion of the diagnosis The external rotation maneuver performed on the x ray table to procure the lateral view should be gently and not forcibly done. The patient is then placed in traction in abduction with an internal rotation strap added to his thigh. A considerable improvement in the range of hip motion can often be obtained by traction prior to operation.

Slipped epiphysis of minimal degree may be treated conservatively by a plaster spica and immobilization of the hip until the epiphyseal line is obliterated. The immobilization should be interrupted by periods of traction to maintain good hip motion

Internal fixation *in situ* is generally felt to be preferable There is a generally accepted rule that displacements of one centimeter or less can be internally fixed *in situ* A minimal slip of this type is compatible with good function of the hip if no further slip occurs.

Fixation *in situ* is done through a lateral proximal femoral incision in a manner similar to that used in treating transcervical fractures of the hip in the adult. A casette is placed beneath the hip and a second portable x ray machine placed under the contralateral thigh and aimed to take a lateral view of the neck with the anterior-superior spine of the ilium in its direct line of the rays. The first machine is moved over the hip for the anterior posterior view when necessary Accurate and firm fixation with the device placed well into the femoral head is necessary

If a nail is used of the tri flanged Smith Peterson type, it should be especially sharpened at its point and the possibility of displacement of the head in the process of driving in the nail borne in mind. If a screw is used the head should be fixed by an external wire in the process of insertion so that rotation of the head does not result in further displacement. It is possible that the im

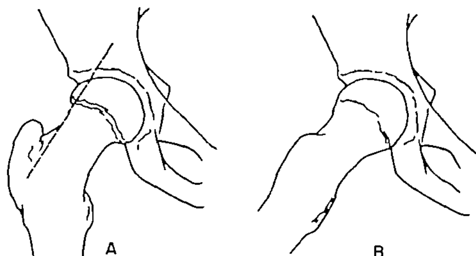


Figure 114 Diagram of the normal relation of the capital femoral epiphysis to the extension of a line drawn along the superior aspect of the neck. A anterior view B lateral view

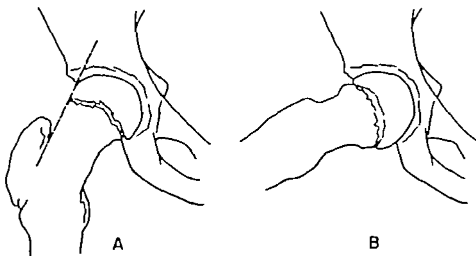


Figure 115 Diagram of slip visible in the antero-posterior view only No slip is seen in the lateral view

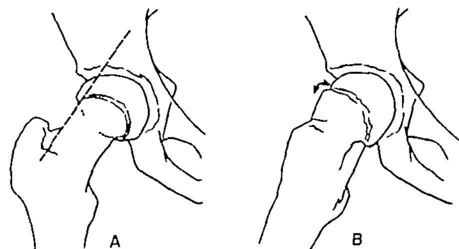


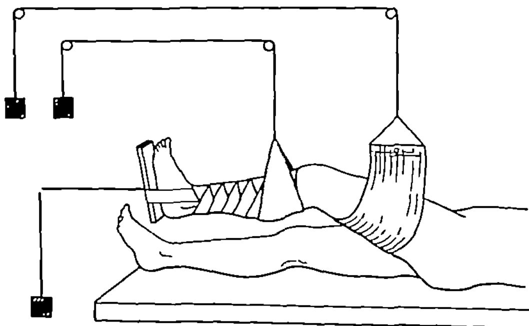
Figure 116 Diagram of slip recognizable in the lateral view only (Figures 114-116 *Redrawn from* Am J Roentgenol 46: 368, 1951)





Figure 117 Slipped epiphysis AP and lateral roentgenograms with displacement visible in lateral view

Figure 118 The use of the internal rotation strap aids in reducing the external rotation contracture The adhesive is protected from pulling on the lateral side of the leg by placing felt between it and the skin.



TRACTION FOR SLIPPED EPIPHYSIS WITH USE OF INTERNAL ROTATION STRAP

paction produced by a screw at the epiphyseal line will result in earlier fusion. In general, the insertion of one or two screws is preferable to the use of nail or pins.

The use of match stick type grafts placed across the epiphyseal line has been advocated by Howorth to hasten closure of the epiphyseal line.

Approximately one out of four cases of slipped epiphysis are seen which are afflicted by a severe degree of slipping and reposition of the epiphysis in relation to the neck. It is necessary to procure a functioning hip which will not degenerate later into the marked limitation brought on by arthritic changes.

A very few of these have an acute history. There is no rounding of the corners of the neck or build up of callus about the slipped epiphysis by x ray in cases of acute slip. Where these have been associated with acute trauma, it is sometimes possible to manipulate the hip with internal rotation and abduction of the lower extremity. When successful on gentle manipulation, the hip may then be nailed *in situ*. The remainder of the acute slip cases require open arthrotomy, correction of the mechanical derangement, followed by internal fixation.

The hip may be approached anteriorly through the Smith Peterson incision as advocated by Klein or posteriorly through a Gibson incision.

The method of correcting the displacement of the epiphysis may be by a wedge osteotomy of the neck. Another method utilizes the area of the epiphyseal line. This area is divided without removing a wedge and the neck slid back into place beneath the epiphysis.

The first method results in shortening of the neck thereby relieving stress on the stretched vessels of the neck. The importance of the retinaculum of Weitbrecht and the maintenance of circulation to the femoral epiphysis by this method has been noted by Green. The second method has been followed by excellent results in the cases treated by Klein *et al*. When a wedge of bone has been removed prior to replacement of the head, an absolute anatomic reduction cannot be reacquired.

The operation, whether through an anterior or posterior approach and with or without the removal of a wedge, has certain underlying principles.

These include gentle handling of the epiphysis and the maintenance of the blood supply to it. Both the internal and external femoral circumflex arteries provide branches running up to the femoral neck. The posterior, superior and inferior portions of the capsule are particularly important. The capsule should be divided parallel with the lip of the acetabulum and next to it. Any further incision, if absolutely necessary, splits the capsule in line with the femoral neck so that as little interference as possible is made with the blood supply.

After the head is divided from the neck, the reduction is carried out with aid of a bone skid and followed by internal fixation from the lateral cortex of the femur.

Post Operative Treatment

The patient is placed in traction until the post-operative discomfort and muscle spasm have subsided. When the hip can be moved freely, the patient

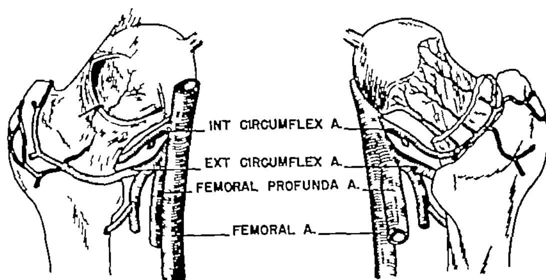


Figure 119 Diagram of the anatomy of the circulation to the proximal femoral epiphysis. Note that transverse division of the capsule proximally (acetabular side) will not jeopardize the circulation

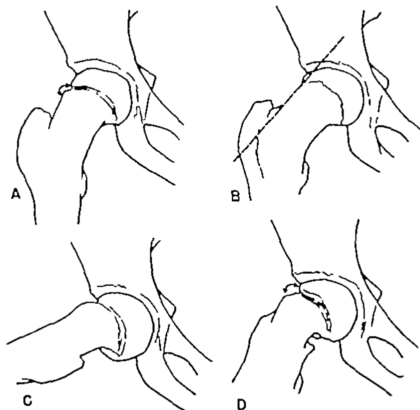


Figure 120 Measurement of the amount of slip is done along the arc of slip as indicated by the arrows in the antero-posterior views (A, B) and lateral views (C D)

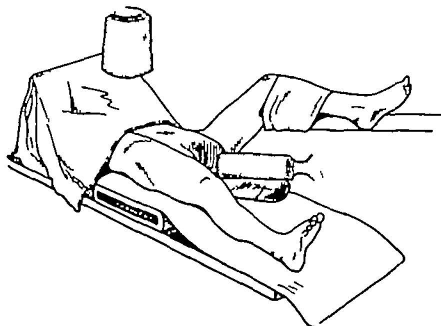


Figure 121 Equipment to procure roentgenograms of the hip is necessary when treating slipped epiphysis operatively. Films must be taken in two planes. The placement of the tube for antero-posterior and lateral views is shown (*Redrawn from* Klein A. *et al.* Slipped Capital Femoral epiphysis. Charles C Thomas Springfield, Illinois, 1953)

is allowed up on partial weight bearing with crutches. Fusion of the epiphysis is noted in two to eight months after open reduction and six months to two years following nailing *in situ*. Once the epiphysis has fused, the internal fixation apparatus is removed.

The possibility of involvement of the contralateral side must be constantly kept in mind and nailing *in situ* performed if the diagnosis is made. Postoperative films should always include both hips. Involvement of the other hip while usually taking place within the first year and one-half after the initial diagnosis may not occur until two or more years later.

The results following nailing *in situ* for minimal slips are excellent. The results following open reduction are good with careful attention to basic requirements in care. This includes sparing the vascular supply to the epiphysis during the operation anatomical repositioning and early mobilization.

The development of aseptic necrosis of the head threatens a successful result. Such complications are rare but occur in all series. Badgeley reports three instances in seventy five hips. The initial displacement may predetermine its development regardless of the method and care used in handling the case.

References

Ferguson A B and Howorth M B Slipping of the upper femoral epiphysis J A. M A 97: 1867 1931

Howorth M. B Slipping of the upper femoral epiphysis. J Bone & Joint Surg., 31A: 734, 1949

Ghormley R. K. and Fairchild R. D Diagnosis and treatment of slipped epiphysis. J A. M A., 114: 229 1940

Klein A, Joplin R., Reidy J and Hanelin J Slipped capital femoral epiphysis. Charles C Thomas, Springfield Illinois.
Wilson P D Treatment of slipping of the upper femoral epiphysis with minimal displacement J Bone & Joint Surg 20: 379 1938

SYNOVITIS OF THE HIP*

Acute synovitis of the hip is one of the most common orthopedic conditions seen It is an inflammatory condition of the hip of short duration, relieved by rest It looms in the forefront of every differential diagnosis of childhood limp Although this condition is so common, it seldom has been described It is related to Legg Perthes disease in that it is potentially preliminary to it

A review of the cases admitted to the Children's Hospital (Pittsburgh) over the last four years reveals that the age distribution is very similar to the three- to ten year-old group found in coxa plana Occasionally, a case as old as twelve may be seen. Four out of five cases are males

Acute synovitis of the hip is sometimes so mild that its symptoms attract little attention sometimes it is so severe that it is mistaken for a septic hip joint Occasionally, cases are recognized only as synovitis by roentgenogram which go on to develop the typical changes of coxa plana.

Etiology

Very frequently there is a history of respiratory illness from ten days to three weeks prior to the onset of hip symptoms The relationship of these predominantly streptococcal infections to the inflammatory changes of the hip is not clear When trauma looms large in the history, the presence of free blood in the joint is postulated Occasionally, this has been confirmed by aspiration Despite this occasionally clear implication, it is apparent that no single etiologic agent exists whether or not such a joint is an allergic manifestation is uncertain.

Clinical Features

The patient is typically a male about the age of five Pain usually begins at the knee in the previous twenty four hours The pain tends to become more localized at the hip as the disease progresses Frequently the temperature at the height of the symptoms is as high as 101 , occasionally, 103 However, the leukocytes ordinarily are not increased. The failure of an increase in the leukocytes aids in differentiating the condition from a septic hip when the temperature is high The sedimentation rate may be slightly elevated.

Motion of the hip always is limited There is temporary flexion contracture and limited rotation particularly internal rotation in flexion together with limited abduction. Palpation of the hip joint reveals both anterior and posterior tenderness

Roentgenographic Findings

There are two definite X ray findings The most obvious of these is swelling of the capsular shadows about the hip. The rounded increased prominence

* See footnote p 112



Figure 122 Synovitis of the hip the outline of the capsular shadows are rounded and increased in width on the right. The normal straight outline is seen on the left.

of the capsular shadows about the hip is seen best superiorly and laterally Accompanying this is a minimal widening of the hip joint. Although slight and often found only by measurement still it is commonly seen. Widening of the hip joint space is recognized more easily in the medial inferior portion of the joint. The anteroposterior view of the hip is most instructive. There are no bone changes suggestive of coxa plana.

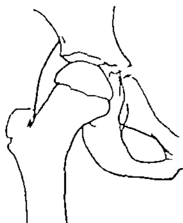
Differential Diagnosis

The hip inflamed because of tuberculous involvement has a chronic history which acute synovitis does not have. Septic involvement of the hip must be differentiated by the more definite signs of sepsis—elevation of the white count and shift to the left. Aspiration may be necessary but in practice rarely turns out to be so. Other possible underlying causes of inflammation of the hip, such as rheumatic fever or rheumatoid arthritis, are evident on general physical examination or future course

Treatment

The inflammation about the hip tends to subside readily when weight bearing is eliminated. Traction in line with the flexion contracture is the most certain





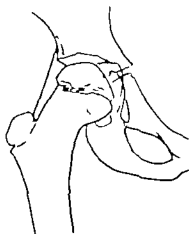




Figure 123 Diagram of the capsular shadows from the accompanying roentgenograms. At top the hip is that of a patient with symptoms and signs of synovitis with bulging capsular shadow Below the hip is normal.

method of obtaining rapid subsidence of symptoms While relief from pain is almost immediate it usually takes from thirty-six to seventy two hours to regain full motion at the hip Failure to regain full motion at the hip within four or five days leads to a suspicion that a more serious etiology underlies the condition

The patient usually is given one week of non weight bearing after full motion has been regained to guard against recurrence. Recurrences lead to a suspicion of coxa plana.

LEGG-PERTHES' DISEASE*

Although synovitis of the hip may be preliminary to coxa plana, aseptic necrosis of the proximal femoral epiphysis may start without any previous difficulty at the hip It is known variously as Legg Perthes disease, osteochondritis deformans coxae juvenilis or coxa plana. The condition may be defined as a disease of the hip limited by age group and largely by sex, it results in changes in the femoral head apparently secondary to a loss of adequate vascular supply Its duration is self limited but its course may result in irreversible mechanical impairment of the hip The age group runs predominantly from three to ten years old with occasionally a case as old as twelve. As in synovitis of the hip the disease occurs predominantly in males (80 per cent) Although Legg Perthes' disease can be bilateral it is most often unilateral. (90 per cent)

Etiology

A good understanding of coxa plana can be achieved if it is considered as a disease of the soft tissues rather than of the bone then the swelling of soft tissue shadows about the hip, present on the roentgenogram, is not lost to observation in the presence of rather striking bone changes Ferguson and Howorth have noted the presence of thickened blood vessel walls and thickened capsule in surgically observed cases They postulated a relationship between previous simple synovitis of the hip or other causes of inflammation at the hip joint and coxa plana. Many have tried to implicate thyroid deficiency because of the occasional occurrence of a similar x ray change at the hip in cretins. Conditions resulting in irregular ossification of the femoral head, such as chondrodysplasia, bear a superficial resemblance by roentgenogram but are not associated with capsular swelling about the hip or with clinical symptoms and usually have other stigmata of the underlying condition It is possible to have an aseptic necrosis of the proximal femoral epiphysis in medical embolic disorders. This includes infections and physiologic variations such as caisson disease.

Aseptic necrosis of the femoral head is seen in patients with congenital dislocation of the hip It has been noted particularly when the hip has been fixed in marked internal rotation. The vascular interference runs a much shorter course than in Legg Perthes disease, apparently because the underlying cause if positional can be relieved more readily

Pathology

The joint very seldom has been observed when involved in the active stages of this disease When it has, thickening of the capsule and the synovium has been found as well as increased thickness of the cartilage of the femoral head and neck. It is felt that this thickening accounts for the widened joint space—an early roentgenographic feature. The synovium may be edematous and hyperemic in stages where motion is markedly limited. The area of metaphysis immediately adjacent to the epiphyseal line may be softened by increased vascularity The outward appearance of the femoral head in early stages does not

* Reprinted by permission from *Clinical Orthopedics* Vol. 4, p 180-188. J B. Lippincott Co Phila., 1954

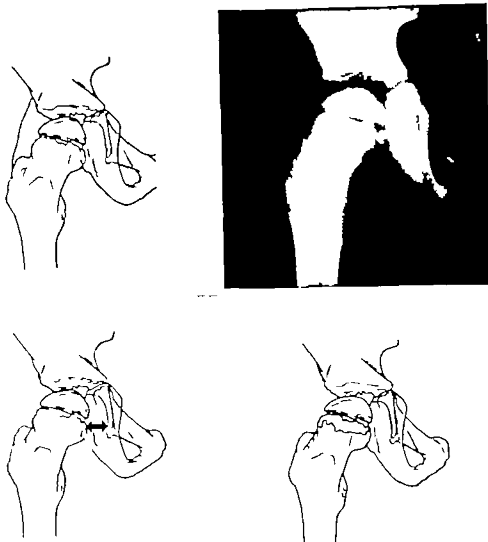


Figure 124 Early signs of Legg Perthes disease of the hip by roentgenogram Bulging capsular shadows, increased density of epiphysis, widened joint space and demineralized metaphysis adjacent to the epiphyseal line are shown

indicate the rather marked changes going on within it. The necrotic bone undergoes a fibrous-tissue replacement process which results in the appearance of radiolucent areas in the dense head shown by roentgenogram. Bony areas remaining may be replaced by creeping substitution. The fibrous areas are replaced by bone but often only after considerable delay

Clinical Picture

Characteristically there is a history of limp and pain at the knee, usually for a period of three weeks to two months before being seen. The patient is usually a boy and most often four to six years old although he may be anywhere in the three- to twelve-year-old group There is a hip flexion contracture readily elicited, and further testing of the motions in flexion develops the limitation of internal rotation and abduction. The hip will come to neutral in rotation There may be tenderness both anteriorly and posteriorly to external palpation of the hip joint Measurement of the mid thigh often reveals atrophy

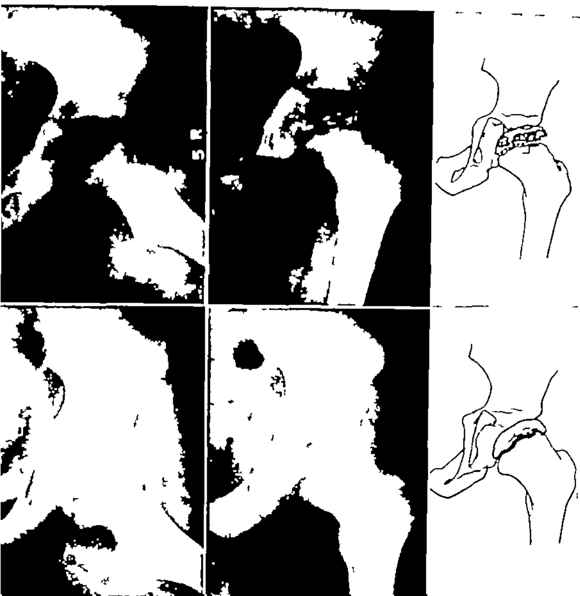


Figure 125 Prediction of expected result in coxa plana from the roentgenogram when first seen. The flattened predicted outline of the head is diagrammed at upper right. The follow-up roentgenogram of the same case and accompanying diagram is below

of one-half to one inch. Even if not measurable atrophy is regularly recognizable by palpation of the thigh and the gluteal area The child may be in acute distress. In contrast with this picture occasionally and rarely there are children who present such mild features that the disease is recognized only by detailed and thorough examination of the motions of the hip Only by taking the time to make a detailed recording of hip motion particularly in flexion, will the examiner avoid the embarrassment of having the case diagnosed more successfully elsewhere.

Not all cases of Legg Perthes disease have symptoms immediately at onset. Some cases first are seen when there is already in existence considerable widening of the neck and the joint space with no history of previous symptoms, then there are changes which are irreversible These are not apparent when the femoral head is radiolucent, but study of the roentgenogram will reveal that the shape or the head can be predicted The stages of coxa plana are discerned best by roentgenogram.

First Stage (Incipient)

There is swelling of the capsular shadows about the hip, widening of the joint space and demineralization of the femoral metaphysis in the neck immediately adjacent to the epiphyseal line.

Second Stage (Aseptic Necrosis)

The changes of the first stage are present, in addition there is definite increase in density of the femoral head in its entirety or in some portion. The area supplied by vascular channels from the ligamentum teres may be spared Often cases are seen first at this stage If no widening of the neck has taken place and the head is outlined fully then no irreversible changes occurred, and treatment has the possibility of restoring a good hip

Third Stage (Regenerative)

Here, revitalization of the head is taking place, as evidenced by the presence of radiolucent areas. Widening of the femoral neck also may be evident. One can outline, on the basis of the neck width and joint space remaining, the area of the head not visualized. Often to do so is illuminating. It may be evident that considerable mechanical malformations still may be compatible with good hip function but the anatomic outline will not be normal. Still further severity in flattening and widening of the head may be present so that a poor result is fore-ordained However treatment should not allow any further progression if it is to be of value. In the last portion of this stage, regeneration of the femoral head takes place until the head is filled in completely

Relation to Coxa Magna

Coxa magna is a frequent end result of coxa plana. Its development can be ascribed to the cartilage overgrowth that apparently takes place under conditions of impaired vascularity at the hip Widening of the femoral neck becomes evident in the second stage of coxa plana when conditions of aseptic necrosis are well established. The regenerating head must match this increased width of neck regardless of whatever other deformity the head may have. The head reformed without deformity other than increased size is consistent with good motion at the hip However such increased width results in emergence of the lateral portion of the head from beneath the overhanging acetabular roof Apparently coxa magna can occur without the development of aseptic necrosis of the head that is as an entity quite separate from coxa plana

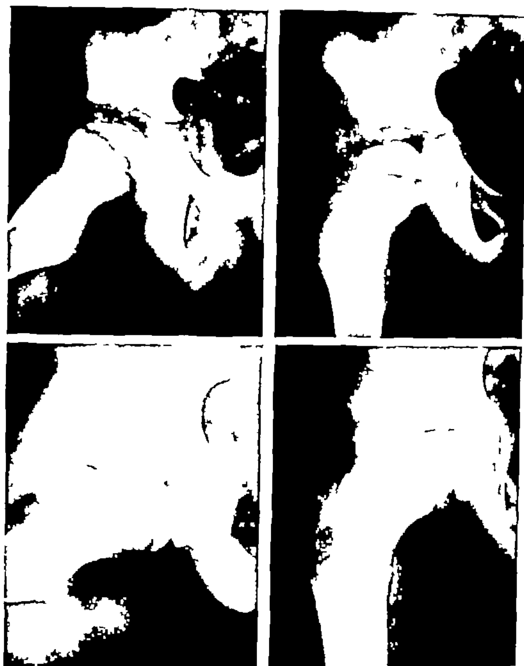


Figure 180 Development of coxa magna from early coxa plana

Treatment

Several authors have noted the importance of bed rest and traction in the treatment of coxa plana. It is helpful to remember that the soft tissues about the hip must be restored to normal before the vascular impairment will cease. Until that time the head must be protected from forces which may cause secondary mechanical impairment of the head by flattening and widening.

Treatment revolves around the maintenance of *full joint motion* a normal



Figure 127 Coxa magna in the adult with degenerative arthritis

femoral head outline and a patient who is a well rounded person, for the disability caused by the late effects of this disease can be a severe deterrent to a person's progress through life.

Some deprivation of activity at the stage of involvement may be necessary to give the patient a useful weight bearing joint in the years ahead. A strict regimen of non weight bearing is essential or there is no sense in advocating this limitation at all.

Removal of the patient from weight bearing is not enough to prevent deformity of the head He must gain relief from muscle spasm. This is accomplished best by traction in line with his flexion deformity It should be continued until there is no limitation of motion It is essential that full motion of the hip be restored regardless of whatever further form of treatment is used. Full motion never should be lost from this point onward. It is obvious that placing the patient in a cast does not fall in line with the previous statement

Should the case be an early one with minimal soft tissue and bony involvement it is possible that the time needed to progress to a regenerative stage may be relatively short These patients regain full motion of the hip quickly and then

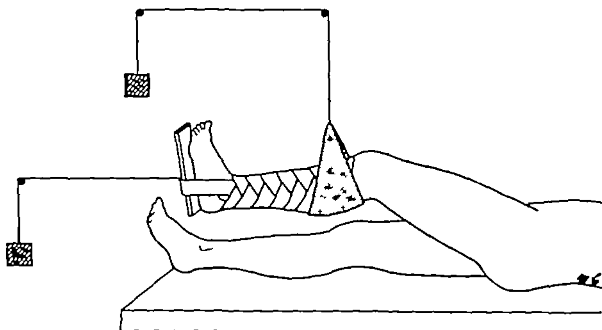


Figure 123 Split Russell traction in line with the flexion contracture of the irritated hip

they may be treated by absolute bed rest with frequent re-examinations to ensure that full motion is being maintained.

In the untreated cases the expected time intervals for each stage are as follows first (incipient) from two to four weeks, second (aseptic necrosis) from *one to two years third (regenerative) from three to four years*

The exact form of achieving non weight bearing is not as important as the fact that it is truly achieved. Goff keeps the child on a rolling platform which allows him to move from place to place. Where the patient has to look forward to a non weight bearing period of considerable duration various means have been devised to allow him to be ambulatory An ischial weight bearing calipes with a patten bottom and the foot suspended three inches from the ground may be used. A corresponding raise on the other shoe is necessary in order to conveniently progress with the splint. Belts and slings holding the knee in flexion have been devised to prevent weight bearing Apparatus which the patient can easily disengage requires extra vigilance from the physician. If the patient cannot be counted on bed rest alone is preferable—often with institutionalization. Whatever method is used the zeal with which it is carried out may save the hip from progressive flattening. Full motion at the hip must be maintained and the patient should be rechecked at two-month intervals at least.

A return to weight bearing is justified when there have been no new dense areas in the femoral head for two months and there are no dense areas in the line of stress trabeculae of the head. The patient must have full motion at the hip joint. There also must be definite evidence that reconstruction is taking place in the head. It is not necessary to wait for a completely remineralized head but sufficient bone structure should be present to arouse confidence in the preservation of that structure



Figure 1[illegible] Legg Perthes disease with patient first presenting late in the disease. Time interval 2 years.



Figure 180 Legg Perthes disease with patient first seen early in the course of the disease. Time interval 1 year

References

EYRE BROOK A I Osteochondritis deformans coxae juvenilis or Perthes disease results of treatment by traction in recumbency J Bone & Joint Surg 24: 166 1936

FERGUSON A B AND HOWORTH M B Coxa magna a condition of the hip related to coxa plana J A M A, 104: 808 1935

FERGUSON A B AND HOWORTH M B Coxa plana and related conditions at the hip J Bone & Joint Surg, 16: 781 1934

LEGG, A T An obscure affection of the hipjoint Boston M & Surg J 162: 202 1910

GOFF CHARLES W Legg-Calve-Perthes syndrome and related osteochondroses of youth Charles C Thomas, Springfield Illinois, 1954

MINDELL, E. R. AND SHERMAN M S Late results in Legg Perthes disease, J Bone & Joint Surg 33A: 1 1951

PEDERSON H E AND MCCARROLL, H R. Treatment in Legg Perthes disease J Bone & Joint Surg, 33A: 591 1951

TUBERCULOSIS OF THE HIP

The onset of this disease is seen as a rule in young children. It forms twenty percent of the cases of bone and joint tuberculosis Involvement of the hip with tuberculosis is commoner than infection of any other single joint.

The infection is hematogenous and due to Koch's Bacillis. The initial lesion may be synovial in the superior acetabulum or in the inferior neck adjacent to the epiphyseal line Since it is impossible to excise the local site of involvement with hope of success, it is best to think of the disease as generally involving the joint.

Clinical Picture

The onset is insidious with a chronic story of limp and hip or knee pain, usually running over many months when first seen Cries in the night with sudden awakening may feature the illness There are systemic complaints often including anorexia loss of weight and afternoon fever The child limps, drops from activities with his fellows and avoids weight bearing.

On physical examination, the atrophy of the limb is often striking The hip motion is limited with a hip flexion contracture evident on flexing the uninvolved hip so that the lumbar spine is flattened. Guarding with pain on attempted motion may be present in all planes but early in the disease may be most evident in internal rotation and abduction as well as extension.

The rate of progression of the disease may vary from individual to individual

The limp is antalgic with a short stance phase on the involved leg. Hip flexion contracture may result in marked lumbar lordosis on standing. Local tenderness of the joint may be evident over the femoral head, located beneath the femoral artery one finger's breadth below the inguinal ligament. Posterior joint tenderness may be present as well

It is not common in these times to see a tuberculous hip accompanied by abscess formation when it is first diagnosed Shortening of the limb from bone destruction is also a late finding Relative shortening may be present due to an adducted position however Some cases late in the disease show *extensive shortening* as much as ten inches. Gill has shown that this may be due in *part to* premature central closure of the distal femoral or proximal tibial epiphysis.



Figure 131 Early tuberculosis of the hip with atrophy and bone involvement

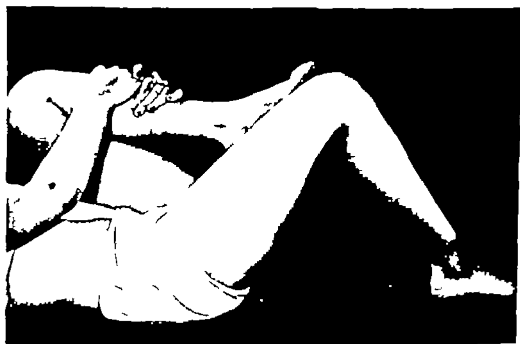


Figure 133 Hip flexion contracture with infection of the hip

Pathology

The disease can begin in the synovia in the metaphysis adjacent to the proximal femoral epiphysis as in the acetabulum. The place of origin is not important. Even if synovial the tuberculous granulation tissue begins invasion of the subchondral area very quickly Metaphyseal foci spread to the same area, but usually not by going through the epiphyseal line A primary epiphyseal foci is uncommon. The trabeculae of the articular cortex are absorbed with granula

tions both over and under the joint cartilage This cartilage may be free of its attachment to the parent bone Areas of contact of joint cartilage constitute a barrier to the extension of the disease and cause destruction to take place first at the joint margins

With the advancement of the disease, complete destruction of cartilage and subchondral bone occurs Lymphocytes and plasma cells infiltrate the granulomatous tissue lying intermixed with fibroblasts and vascular tissues. Typical tuberculous arrangement may be found with epipeloid and giant cells about a central area of caseation In children healing may occur spontaneously but usually with a fibrous ankylosis resulting.

Roentgen Picture

Despite the relative resistance of cartilage to the infection, the most noticeable roentgen finding may be narrowing of the joint space. There is apparently no proteolytic enzyme associated with tuberculous infection, as in pyogenic pus which will digest cartilage. Nonetheless progression of the disease is so slow and chronic that narrowing of the joint space actually stands out.

There is active destruction of bone which proceeds irregularly rather than in rounded distribution as in a tumor both sides of the joint are often affected. Later marked bone atrophy occurs in the involved area. There is distention of the capsular shadow and a hazy loss of distinctness of the trabeculae over a wide area The disease may progress to gross destruction.

Treatment

The first objective is to make the diagnosis of this destructive lesion without doubt. While studies are proceeding the child should be confined to bed with traction at the line of his flexion deformity with the object of reducing the degree of contracture eliminating muscle guarding, and securing comfort for the patient.

There is a positive tuberculin test virtually without exception. A negative test in a patient suspected of having tuberculosis should result in checking the antigen and using another source The laboratory work usually reveals an elevated sedimentation rate and may reveal a leucocytosis The lymphocyte-monocyte ratio may be altered. Secondary anemia may be present.

The joint should be biopsied and inspected. The degree of cartilage involvement in both sides is noted at the time of biopsy With confirmation of the diagnosis one proceeds with treatment.

In joints still having intact surfaces an attempt should be made to save the joint This usually means immobilization in a hip spica which also includes the other leg to the knee. The spica is bivalved and the patient allowed to briefly carry the joint through a range of motion twice daily The spica is not applied until the joint flexion and adduction contracture has been corrected in traction.

The patient's systemic response should subside rapidly if the treatment is to be successful

Chemotherapy using streptomycin, isoniazid and para amino salicylic acid is used as outlined in the treatment of the knee with this disease.

Failure of the systemic symptoms to rapidly subside and *progression of de-*

struction by roentgenogram indicates the need for complete immobilization with the hip in neutral position in relation to the pelvis.

Such treatment is also indicated if the patient first presents with a destroyed joint which cannot be functional. Abscesses of the hip are drained and closed primarily

In those cases destined for fusion, the systemic symptoms are first brought under control. The younger the patient the more difficult it is to secure fusion, and the longer time it is delayed. Below the age of ten fusion is frequently inadvisable

Attempts at fusion directly through the area involved by disease do not do well and subjects the grafts to the ravages of the tuberculous granulation tissue. Operations for hip fusion try in general to by pass the diseased area either above or below the hip joint.

In Wilson's technique a rectangular section of ilium is turned down into the trochanter In Hibb s technique the greater trochanter and a portion of the shaft is cut out and reversed and the shaft end is sunk into a hole in the ilium, the cortex of which has been levered out The cut surface of the graft faces the neck which can be denuded where healthy All fusions superiorly have one defect in common the muscles guarding the hips are tending to pull it into adduction, thus separating the graft from bony contact Thus some force tends to compress into the graft when it is inferior Brittain s method runs the graft from the femoral to the ischial region A tibial graft is run through a subtrochanteric osteotomy of the femur into an ischial bed. In the Trumbell operation the tibial graft runs from femoral shaft to ischium but without a subtrochanteric osteotomy

In all hip fusion operations the task is easier if hip motion is limited and the joint quite firmly fixed if unstable difficulty in fusion may be encountered.

All of these opreatons require immobilization in a bilateral hip spica for at least six months Thereafter release from immobilization depends on the state of bone fusion Bony union takes place last through the diseased area The by pass graft acts as a stabilizing start to eliminate motion and permit healing of the diseased area.

References

Bosworth D M Wright H A and Fielding, J W Treatment of bone and joint tuberculosis effect of (1) isonicotinyl (2) isopropylhydrazine J Bone & Joint Surg 34A: 761 1952

Bosworth D M Wright H A, Fielding J W and Wilson H. J Jr. Use of iproniazid in treatment of bone and joint tuberculosis. J Bone & Joint Surg 35A: 557 1953.

Gill, S D The cause of discrepancy in length of the limbs following tuberculosis of the hip in children. J Bone & Joint Surg 26: 272 1944

Mercer, W Management of the tuberculous hip joint. J Bone & Joint Surg 36A: 1123 1954.

PYOGENIC INFECTION OF THE HIP

Infected hip joints are seen most frequently in infancy and may easily be overlooked in the early stages. Early diagnosis is necessary to save the hip joint. It occurs most often as a complication of a general septicemia. The initial

lesion may be respiratory, skin, enteric, or an umbilical vein infection It is not unknown as a complication of femoral vein puncture in a septicemic infant

The disease may begin as an osteomyelitis of the femoral neck metaphysis rather than a primary synovial infection The femoral neck being contained within the joint rather easily allows progression of the infection directly into the joint due to its peculiar anatomy Destruction of cartilage is particularly possible with staphylococcic infection due to its proteolytic enzyme, a fact which has been pointed out by Phemister

Clinical Picture

The patients fall into two groups those who are acutely ill and appear so, and those who are acutely ill and do not appear so Fever and a high leucocyte count although helpful do not have to be present to make the diagnosis in infancy Failure to move the limb may be the first clinical observation Swelling in the hip and trochanteric area may be another Some possible portal of entry for the organism is usually known. The most common agents are the streptococcus staphylococcus, pneumococcus or *Bacillus coli.*

Examination reveals limited motion of the hip This is most easily noted in both internal and external rotation—no rotation being evident Swelling obscures bony prominences and makes palpation of the femoral head beneath the femoral artery difficult or impossible Tenderness is present both anteriorly and posteriorly over the hip joint.

Roentgen Picture

The early signs are those of distension of the capsular shadows of the hip joint and widening of the joint space Later, areas of destruction in the metaphysis of the femoral neck may be seen. If the infecting agent is the staphylococcus bony proliferation may also be noted relatively early In infants with so much of the femoral epiphysis and neck consisting of cartilage, the amount of destruction may not be evident.

In the untreated case further distension of the joint with purulent material plus the associated muscle spasm leads to dislocation. Subperiosteal new bone may be present well down the diaphysis in late cases

Treatment

The disease is an emergency since drainage and removal of purulent material in contact with the cartilage of the femoral epiphysis may be the only means of saving it from destruction. The general condition of the patient may be reenforced by transfusion before proceeding if necessary

On clinical suspicion the joint is aspirated anteriorly with a number eighteen spinal needle The purulent material will well up into and out of the needle on entering the distended hip joint. Smears and cultures of this material are made

If pus is obtained the patient is turned over and the buttock area reprepared and redraped. Drainage of the hip is done through a linear Ober type incision. This incision is made in line with the femoral neck and over it. The landmarks are a point one third of the distance along the iliac crest from the posterior su

Figure 153 Soft tissue inflammatory swelling widening of the joint space and displacement of the femoral shaft at the hip in acute pyogenic infections of the hip

Figure 154 Actual dislocation resulting from hip joint infection



Figure 135 Hip flexion and adduction with fixed deformity following previous sepsis of the hip joint.

perior spine and the trochanter of the femur In infants, care should be taken to keep the incision well lateral to avoid the sciatic nerve. The incision splits the fibres of the gluteus maximus, separates the quadratus femoris from the gemellus and then incises the hip joint capsule The joint is thoroughly irrigated so that all fibrin clots are removed. A suitable chemotherapeutic agent is instilled. The edges of the capsule are sewn open with a few catgut sutures. A catheter is led down to this opening but not into the joint itself and held in place by a skin silk. The wound falls together and the skin is loosely approximated

Postoperatively the patient is treated on a Bradford frame. The posterior incision allows for gravity drainage and does not interfere with nursing care. The catheter is gradually removed over the next five days with variation depending on the patient's clinical course.

During therapy and from the time of recognition, the patient has both legs in traction preventing dislocation and allowing for the resumption of early motion. Immobilization in a hip spica during this period is too likely to lead to a limited hip motion

References

DUNHAM E. C Septicemia in the newborn. Am J Dis. Child., 45: 220 1933
FRASER, J Discussion on acute osteomyelitis. Brit. M J., 2: 605 1924
POTTER, C M C Osteomyelitis in the new-born. J Bone & Joint Surg 36B: 578 1954.

4

The Spine

CONGENITAL KYPHOSIS

An important childhood orthopedic problem is kyphosis of the spine based either on a widespread disturbance of ossification of the spine or a localized vertebral anomaly

The patient is seen because of an increased antero-posterior curvature resulting in a rather sharp prominence usually centering about the twelfth dorsal vertebrae.

Etiology

There are at least three conditions in which this lesion is frequently seen. Widespread disturbance of ossification of the spine is often seen in chondrodystrophy Such spines frequently have a typical shape of the individual vertebra where the anterior superior and inferior angle is not ossified. Gargoylism and cretinism frequently have associated with them a hyper mobility at the twelfth dorsal first lumbar area. The twelfth dorsal is often wedge-shaped and irregular

Individual vertebral anomalies of the type producing kyphosis are most often *centered at the junction of the dorsal and lumbar spine. Such anomalies consist* of various partial segments of the vertebra involved the posterior portion being present, the anterior absent.

Clinical Picture

Morquio's Disease

Morquio's disease is a syndrome constituting the form of chondro-dystrophy in which the changes are most prominent in the spine It often is not recognized until the child is walking. The trunk is short, out of proportion to the shortening of the extremities. *Skull* changes are minimal *The kyphosis is diffuse with* wedging of the vertebrae in addition to localized changes at the twelfth dorsal. The extremities reveal widened epiphyses and bizarre bone shapes Bone that is laid down has normal texture however There may be associated deformities, such as knock knees. There is an epiphyseal disturbance of the vertebrae as well as extremities The epiphyseal line reveals a lack of cartilage column formation



Figure 137 Lateral roentgenogram of the spine in a cretin revealing the hypermobility between the twelfth dorsal and first lumbar area.

4

The Spine

CONGENITAL KYPHOSIS

An important childhood orthopedic problem is kyphosis of the spine based either on a widespread disturbance of ossification of the spine or a localized vertebral anomaly

The patient is seen because of an increased antero-posterior curvature result ing in a rather sharp prominence usually centering about the twelfth dorsal vertebrae.

Etiology

There are at least three conditions in which this lesion is frequently seen. Widespread disturbance of ossification of the spine is often seen in chondrodystrophy Such spines frequently have a typical shape of the individual vertebra where the anterior superior and inferior angle is not ossified. Gargoylism and cretinism frequently have associated with them a hyper mobility at the twelfth dorsal first lumbar area. The twelfth dorsal is often wedge-shaped and irregular

Individual vertebral anomalies of the type producing kyphosis are most often centered at the junction of the dorsal and lumbar spine Such anomalies consist of various partial segments of the vertebra involved, the posterior portion being present, the anterior absent.

Clinical Picture

Morquio's Disease

Morquio s disease is a syndrome constituting the form of chondro-dystrophy in which the changes are most prominent in the spine It often is not recognized until the child is walking. The trunk is short, out of proportion to the shortening of the extremities Skull changes are minimal. The kyphosis is diffuse with wedging of the vertebrae in addition to localized changes at the twelfth dorsal. The extremities reveal widened epiphyses and bizarre bone shapes Bone that is laid down has normal texture however There may be associated deformities, such as knock knees. There is an epiphyseal disturbance of the vertebrae as well as extremities The epiphyseal line reveals a lack of cartilage column formation



Figure 13C Lateral roentgenogram of the spine in a cretin revealing the hypermobility between the twelfth dorsal and first lumbar area.

4

The Spine

CONGENITAL KYPHOSIS

An important childhood orthopedic problem is kyphosis of the spine based either on a widespread disturbance of ossification of the spine or a localized vertebral anomaly

The patient is seen because of an increased antero-posterior curvature result ing in a rather sharp prominence usually centering about the twelfth dorsal vertebrae.

Etiology

There are at least three conditions in which this lesion is frequently seen. Widespread disturbance of ossification of the spine is often seen in chondro-dystrophy Such spines frequently have a typical shape of the individual vertebra where the anterior superior and inferior angle is not ossified. Gargoylism and cretinism frequently have associated with them a hyper mobility at the twelfth dorsal first lumbar area The twelfth dorsal is often wedge-shaped and irregular

Individual vertebral anomalies of the type producing kyphosis are most often centered at the junction of the dorsal and lumbar spine Such anomalies consist of various partial segments of the vertebra involved the posterior portion being present, the anterior absent.

Clinical Picture

Morquio's Disease

Morquio s disease is a syndrome constituting the form of chondro-dystrophy in which the changes are most prominent in the spine. It often is not recognized until the child is walking. The trunk is short, out of proportion to the shortening of the extremities. Skull changes are minimal. The kyphosis is diffuse with wedging of the vertebrae in addition to localized changes at the twelfth dorsal. The extremities reveal widened epiphyses and bizarre bone shapes. Bone that is laid down has normal texture however There may be associated deformities, such as knock knees. There is an epiphyseal disturbance of the vertebrae as well as extremities The epiphyseal line reveals a lack of cartilage column formation

are found in this condition Hurler's syndrome is associated with mental deficiency and clouding of the cornea The hands sometimes exhibit an inability to extend the distal phalanges resulting in a claw like deformity

Kyphosis Associated with Single Areas of Bony Anomaly

Kyphosis associated with single areas of bony anomaly is usually associated with a *sharper kyphosis* and normal mentality It results in slight diminution in trunk height. Neurological involvement has been reported in one quarter of the published cases according to Bingold.

The most common site of involvement lies between the tenth thoracic and the second lumbar *The type of anomaly varies from the complete absence to presence* of some portion of the posterior elements or body

Treatment

Considerable time and effort has been spent in many orthopedic clinics trying to maintain the patient's spine in a corrected position. Plaster shells, turnbuckle antero-posterior wedging jackets and simple body casts and braces are all used.

In cases where a considerable change takes place in the kyphos in flexion and extension in the upright posture, holding the spine in a corrected position has some merit Such a condition frequently exists in various forms of chondrodystrophy In cases of a single anomaly where the occiput is well centered over the pelvis, the follow up films of such cases indicate that while the condition enlarges with growth the angle of the deformity remains the same

Attempts to reduce the cosmetic deformity by excising the spines are usually misguided. The spines of the involved vertebrae are often rudimentary with flat laminae enclosing the dural sac

The consideration of spinal fusion as a means of maintaining the spine in a position of maximum extension through the kyphotic area is a serious one. *Growth anteriorly in the presence of fused posterior elements may result in* further correction. The posterior elements may be congenitally fused in localized anomalies of the spine. The type of congenital kyphosis most likely to benefit from fusion is that associated with excessive mobility at the area of kyphosis The outlook in syndromes such as Hurler's syndrome with its associated mental deficiency must be considered in making a decision for surgery

References

BINGOLD A C Congenital Kyphosis. J Bone & Joint Surg., 35B: 579 1953
LOMBARD P AND LEGENISSEL, Kyphose congenitale Rev d'Orthop 25: 532 1938

VERTEBRAL PLANA (CALVÉ'S DISEASE)

Vertebral plana characterized by a flattening of the vertebral body which gives a striking X ray picture was first reported by Calvé in 1924 It occurs most frequently between four and ten years but is occasionally seen in the early teens" The original etiology postulated by Calvé was that this represented aseptic necrosis—a process similar to coxa plana. More recently (1954) Com

and as a corollary a lack of provisional zone of calcification. There is no disturbance of membranous ossification

The hand in chondrodystrophy is broad with short fingers and a tendency for the fingers, when outstretched, to diverge, not at the metacarpophalangeal junction, but at the proximal interphalangeal joints.

Hurler's Syndrome

Hurler's syndrome, also known as gargoylism and dysostosis multiplex, resembles Morquio's disease closely. Many of the features of chondro-dystrophy

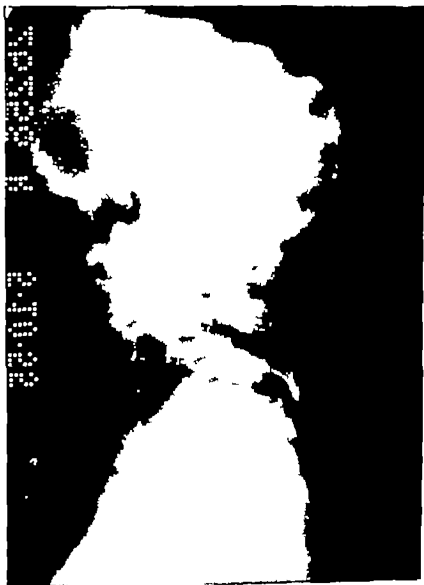


Figure 137 The spine in chondrodystrophy with kyphos at the twelfth dorsal. The *antero-posterior diameter of the vertebral body* is increased and ossification is deficient at the anterior superior and inferior angles of some vertebrae

are found in this condition Hurler's syndrome is associated with mental deficiency and clouding of the cornea The hands sometimes exhibit an inability to extend the distal phalanges, resulting in a claw like deformity

Kyphosis Associated with Single Areas of Bony Anomaly

Kyphosis associated with single areas of bony anomaly is usually associated with a sharper kyphosis and normal mentality It results in slight diminution in trunk height Neurological involvement has been reported in one quarter of the published cases according to Bingold

The most common site of involvement lies between the tenth thoracic and the second lumbar The type of anomaly varies from the complete absence to presence of some portion of the posterior elements or body

Treatment

Considerable time and effort has been spent in many orthopedic clinics trying to maintain the patient's spine in a corrected position. Plaster shells, turnbuckle antero-posterior wedging jackets and simple body casts and braces are all used.

In cases where a considerable change takes place in the kyphos in flexion and extension in the upright posture, holding the spine in a corrected position has some merit Such a condition frequently exists in various forms of chondrodystrophy In cases of a single anomaly where the occiput is well centered over the pelvis the follow up films of such cases indicate that while the condition enlarges with growth the angle of the deformity remains the same

Attempts to reduce the cosmetic deformity by excising the spines are usually misguided. The spines of the involved vertebrae are often rudimentary with flat laminae enclosing the dural sac.

The consideration of spinal fusion as a means of maintaining the spine in a position of maximum extension through the kyphotic area is a serious one Growth anteriorly in the presence of fused posterior elements may result in further correction. The posterior elements may be congenitally fused in localized anomalies of the spine. The type of congenital kyphosis most likely to benefit from fusion is that associated with excessive mobility at the area of kyphosis The outlook in syndromes such as Hurler's syndrome with its associated mental deficiency must be considered in making a decision for surgery

References

Bingold A C Congenital Kyphosis. J Bone & Joint Surg 35B: 579 1953
Lombard P and LeGeniesel, Kyphosis congenitale Rev d'Orthop 25: 532 1938

VERTEBRAL PLANA (CALVÉ'S DISEASE)

Vertebral plana characterized by a flattening of the vertebral body which gives a striking X ray picture, was first reported by Calvé in 1924. It occurs most frequently between four and ten years but is occasionally seen in the early teens The original etiology postulated by Calvé was that this represented aseptic necrosis—a process similar to coxa plana More recently (1954) Com

pore *et al* have presented four cases which were diagnosed as eosinophilic granuloma. Similar deformities have been noted in patients who have widespread areas of involvement with eosinophilic granuloma.

Clinical Picture

The usual complaint is back pain. There is no radiation ordinarily The symptoms are of short duration. If progression has taken place, a gibbus may be found with muscle spasm and limited spine motion.

X ray Picture

When seen early the process may be osteolytic gradually going on to the production of the thin, collapsed "wafer like' body which has been felt to be characteristic of this disease This is quite a different picture from tuberculosis.

Calvé noted that the discs adjacent to the involved vertebrae were intact, that ordinarily only one vertebra was involved, and that the flattened vertebra remained as a dense cortical disc

Treatment

Rest in recumbency during the acute stage of symptoms followed by a spine brace until the lesion is well healed and recalcified is the usual program. Following this the brace is gradually discarded. The usual course varies between one and three years.

References

Calvé, Jacques A localized affection of the spine suggestive of osteochondritis of the vertebral body with the clinical aspect of Pott s disease J Bone & Joint Surg., 7: 41 1925.
Compere, E. W Johnson W E. and Coventry M B Vertebral plana (Calvé's disease) due to eosinophilic granuloma. J Bone & Joint Surg 36A: 969 1954

SPONDYLOLISTHESIS

Spondylolisthesis a low back condition although most frequently noted as symptomatic in the age group from thirty to forty years is also often seen in childhood.

The essential part of this lesion is a defect in the pars interarticularis of the involved vertebra at the junction of pedicle and lamina (Spondylolysis) When bilateral the body pedicles and superior articular processes are separated from the laminae spine and inferior articular processes (Spondyloschisis) This situation may allow a forward slip of the body (Spondylolisthesis)

The most commonly involved vertebra is the fifth lumbar but rarely the fourth lumbar may be involved

Etiology

Ossification of the vertebra springs from three centers—one for the body and one for each half of the neural arch. Ossification starts in the pars interarticularis where the vessels originally penetrate the cartilage anlage.

Willis noted the spondylolisthesis defect" in six per cent of dissecting room subjects with twenty five per cent of these being unilateral In no reported in-



Figure 138. Vertebral plana with preservation of the disc spaces but reduction of the single involved vertebra to wafer-like thinness.

Figure 139. Cleared specimen of lumbo-sacral spine from the newborn. The dark ossification centers of the posterior elements of the lower three lumbar vertebrae are seen coming down to the sacral spine and a portion of each ilium. The isthmus is the point of entrance of the vessels at junction of pedicle and lamina. There is only one center on each side for the posterior elements but the isthmus is the thinnest part of it.

stance has any evidence of bone repair been found even in the unilateral type. Hitchcock notes that despite a five per cent incidence in the population at large none has been found in foetal material. He found in stillborn and infant cadavers that hyperflexion of the spine fractured the neural arch in the lower lumbar region, and tended to support fracture as the basic etiology. A congenital defect is supported by a higher incidence of associated defects such as spina bifida in the presence of the lesion. The argument between the two points of view is still open.

A third possibility exists, however, in that absorption may exceed accretion at the pars interarticularis which would account for the failure of the lesion to be found in new borns as in a congenital defect and the lack of bone repair as in a fracture. Such a theory would account for the high incidence in a group whose occupaitonal habits include prolonged flexion of the spine, but it is also unproven.

Clinical Picture

The patients presenting in childhood are usually first seen in early adolescence. The male and female population appear to be equally involved. Low back pain is the principal complaint with occasional radiation to both thighs posteriorly. Unilateral leg pain seen in one third of the cases in adults is rare in children. The low back pain is made worse by activity and relieved by recumbency.



Figure 140 Spondylolisthesis with defect posterior to pedicle. The fifth lumbar has slipped forward and there is cupping of the sacrum around the posterior inferior angle of the fifth lumbar.

Hamstring spasm limiting straight leg raising is often present and symmetrical Forward flexion of the spine is limited and painful if the child is having symptoms Excessive forward flexion has been found by us only in children who are asymptomatic and where spondylolisthesis was only an incidental finding Tenderness over the spine of the involved vertebra is almost invariably present and localized The trunk, on examining the patient from the lateral view, appears to be displaced forward in relation to the pelvis

Roentgen Findings

Forward displacement of the fifth lumbar on the sacrum can be recognized in minimal cases by drawing a line between the inferior angle of the first sacral and the superior angle of the fifth lumbar A second line is drawn between the superior and the first sacral and the inferior angle of the fifth lumbar as illustrated These two lines should normally be parallel. Forward slip is indicated by a deviation of these lines

The defect in the pars interarticularis may be seen in either the lateral or antero-posterior views of the lumbo-sacral joint. When not visualized, oblique views or a forty five degree antero-posterior view of the lumbo-sacral joint will flatten out the lamina so that it may be visualized, if present

In long standing spondylolisthesis the antero-posterior view is often diag-



Figure 141 Marked atrophy of the laminae of the fifth lumbar vertebra in spondylolisthesis

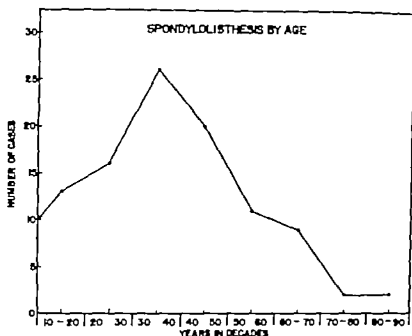


Figure 142 A series of symptomatic spondylolisthesis illustrating age when symptoms bring the patient to medical attention.

nostic since there tends to be considerable atrophy of the laminae involved. A concavity in the inferior surface of the fifth lumbar vertebra may exist apparently due to excessive lumbo-sacral motion The disc space may be narrowed over the normal Spina bifida of the neural arch is frequently seen.

Treatment

Gill has recommended excision of an overgrowth of fibrocartilaginous tissue present beneath the defect and intruding on the fifth lumbar root. He has demonstrated this in his cases but we have found fusion necessary to relieve symptoms in addition to relieving root pressure from whatever cause

In general and depending on the individual patient the attitude has been taken that the basic cause of the symptoms is lumbo-sacral instability and motion of the detached fragment directly over the roots

A lumbo-sacral joint weak enough to be giving symptoms in the presence of the healthy musculature of childhood will almost certainly cause adult disability

Excision of the spine and laminae and fusion of the first sacral to the fifth lumbar vertebrae as recommended by Barr has been carried out in our cases. This has resulted in relief of pain, elimination of hamstring spasm and good spine motion. The area between the fourth lumbar and first sacral is bridged by a continous iliac graft on which bone chips are laid so that there is no danger of a repairative reaction involving the roots. Such cases are ordinarily kept in a double hip spica for four months

Asymptomatic cases are followed but not operated on unless they become symptomatic Strangely enough, mild degrees of slip seem more productive of symptoms than extensive slips involving the total vertebral surface

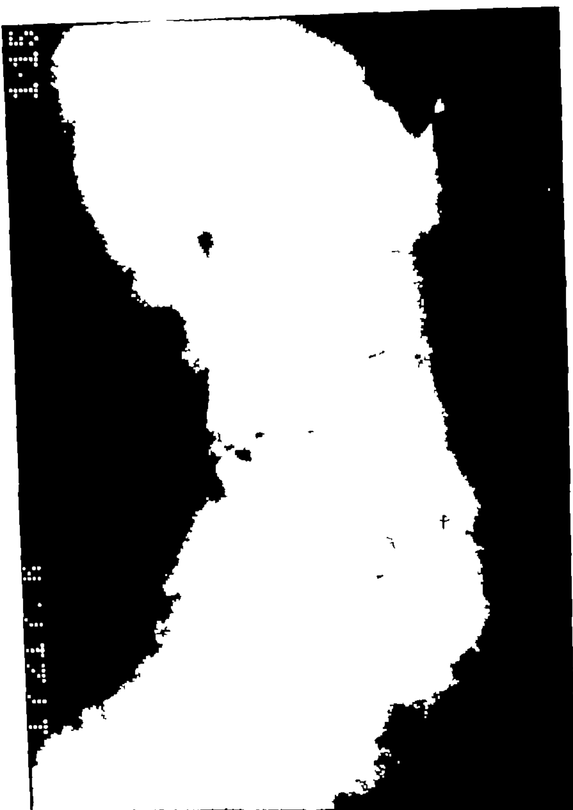


Figure 143 Severe spondylolisthesis slipping forward the entire width of the sacrum



Figure 144 Post-operative view of same case as Figure 143, with partial reduction of the slip removal of the posterior element and fusion posteriorly

References

BATTS M The etiology of spondylolisthesis. J Bone & Joint Surg 21: 879 1939

CALDWELL, G A. Spondylolisthesis, analysis of 89 cases Ann. Surg 119: 485 1944.

GILL, G G MANNING J S AND WHITE, H L. Surgical treatment of spondylolisthesis without spine fusion, excision of the loose lamina with decompression of the nerve roots. J Bone & Joint Surg 37A: 493 1955

HITCHCOCK, H H. Spondylolisthesis observations on its development, progression, and genesis. J Bone & Joint Surg 22: 1 1940

FRIBERG, STEN Studies on spondylolisthesis. Acta Chir Scandinavica, 82: supplementum 55 1939

ROCHE, M B AND BRYAN E S Spondylolisthesis. Additional variations in anomalies in the pars interarticularis. Arch. Surg 53: 675 1946

SHORE, L. R. Report on a specimen of spondylolisthesis found in the skeleton of a Bantu native of South Africa. With further specimens illustrating an anomalous mode of development of the lower lumbar vertebrae Brit. J Surg 16: 431 1929

TUBERCULOSIS OF THE SPINE

Tuberculosis of the spine, while decreased in incidence, remains a condition which is frequently found on orthopedic children's wards It continues to form a formidable percentage of all bone and joint lesions

Patients, in former times, were seen with well advanced lesions The earlier stages in which they are now first seen demands a clinical awareness on the part of the examining physician It is not just a disease of one localized spine area, but must at least in the past have been a generalized infection Even the spine involvement is often multiple. The organism has been blood borne at some stage in order to secure the spine lesion.

Potts first described the disease in 1779 after examination of patients in whom the disease was well advanced and the deformity marked He noted that, by history, symptoms in the spine preceded the development of the deformity

Etiology

Now that the incidence of bovine tuberculosis has been so greatly reduced, the great majority of infections of bone appear to be caused by the human form of Koch s bacillus Spread of the disease is via respiratory rather than alimentary infection It is most usual to find that the patient has had a close family association with an infectious individual. Although the possibility of infection via the thoracic duct to the dorsal spine has been raised, it appears that a blood borne infection is more likely and borne out by the initial localization in the vertebra A primary lung complex found on a roentgenogram of the lungs is common in child hood.

Pathology

The most usual involvement is central in the area supplied by the posterior spinal artery From there the lesion spreads to the disc edge and involvement of the disc Spread includes the anterior and posterior ligaments, but tends to spare the posterior elements including transverse processes, spines and facets

Doub and Badgeley note three types of lesions intervertebral, central and anterior The rare anterior localization is apparently via branches of the intercostal arteries and involves a large number of vertebrae, but often with little deformity

There may be several or many discrete lesions in the vertebra rather than mere direct extension. The initial lesion following acute inflammation and lymphocytic infiltration goes on to central degeneration caseation and calcification. Increasing destruction of bone is produced with further debris and caseation and eventually the formation of an abscess Dorsal abscesses tend to run along the ribs lumbar abscesses to run down the psoas sheath and point either posteriorly in the lumbar triangle or anteriorly beneath and distal to the inguinal ligament. Cervical lesions usually have the abscess gravitating into the mediastinum.

Cave reporting on a series of one hundred and sixty two cases, noted seven in the cervical spine and one hundred and fifteen in the dorsal spine. The dorsal spine is by far the commonest site In this area the normal dorsal round back ac



Figure 145 Tuberculosis of the spine at the dorso-lumbar junction with destruction in opposing vertebrae collapse and deformity without marked calcific reaction.

centuates anterior stress and leads to deformity as contrasted to the cervical and lumbar areas

The majority of initial infections of the spine occur between the ages of three and ten although the disease also occurs in adults.

Clinical Picture

The symptoms are led by pain and stiffness in the back. The parents note wasting loss of appetite night cries withdrawal from activity and easy fatigue. A low grade temperature elevation frequently accompanies the disease. A chronic slowly developing course is typical



Figure 146 Soft tissue shadow of abscess on left with minimal evidence of vertebral involvement.

Figure 147 Gibbus in dorsal spine marking the early devel opment of deformity in the dorsal spine.



The sign which immediately raises the question of a tuberculous infection of the spine is a gibbus This localized prominence in the back is tender to pressure and may only be minimal to observation. There is accompanying muscle spasm with limitation of motion of the spine in all planes. Patients seen before the development of a gibbus exhibit dislike of jarring activity such as jumping and usually some limitation of motion may be elicited. Further progression of the disease results in kyphos in the dorsal spine shortening of trunk height and descent of the ribs into the pelvis.

Abscesses radiating from the lumbar spine down the course of the ilio-psoas may be palpable posteriorly in the ilio-lumbar area anteriorly in the thigh or anteriorly by abdominal examination Abscesses and infections of the rib may be noted Abscesses may be accompanied by flexion contracture of the hip

Laboratory Findings

The sedimentation rate is elevated and becomes of value in following the patients course rather than for diagnosis. The tuberculin test is positive and should first be used in dilution of 1 100 000 These at 1 10,000 are virtually always positive. Failure to obtain a positive test in a suspicious case should result in re-evaluation of the tuberculin being used. The blood count may be unremarkable, but some authors have drawn attention to the lymphocyte-monocyte ratio A rise in monocytes relative to lymphocytes above a ratio of one to three correlates well with activity of the disease and is one additional guide post to the appropriate time for surgery, if indicated.

Roentgen Findings

In the usual stage at which a patient is first seen in this era, narrowing of the intervertebral disc area is an outstanding sign. Active destruction may be found often in the vertebrae both above and below the narrowed interval between. When these findings are coupled with the visualization of a soft tissue mass adjacent to the involved area the diagnosis of tuberculosis becomes most likely Such a soft tissue mass should show a gravity effect, that is the more distal portion of the mass should be larger, gradually narrowing in a cephalad direction. Obviously granulomatous masses will show this effect less than a mass composed of encased fluid pus

Tuberculosis for the chronicity of the lesion exhibits very little calcifying reaction as compared to a pyogenic spondylitis. Organisms such as brucella may mimic the picture very well, however

Late stages of the disease may exhibit such complete destruction of vertebral bodies that deformity, particularly dorsal kyphos is marked and a clear recognition of structure in the involved area unobtainable. Abscess shadows may become especially prominent. In the lumbar area the ilio-psoas shadow may be obliterated by the developing abscess. The entire spine should be x rayed as unsuspected silent lesions may be found.

PARAPLEGIA

This distressing complication of tuberculosis of the spine appears to originate in extradural pressure due to abscess formation. It is most likely to occur with mid-dorsal lesions. Bony deformity, although undoubtedly contributing to the mechanics of the complication, has not been implicated to the degree that granulation tissue debris and purulent material have. Such material pressing on the cord as a tight abscess sac has resulted in the slow development of paraplegia in 23 9 per cent of cases in a series reported by Bosworth Dell Pietra and Rahilly They noted that small abscesses tightly bound down by ligaments were of greater danger than large unconfined sacs

Sustained clonus is a most reliable sign in the presence of a known tuberculous spine lesion. It may be accompanied by sensory changes, muscular weakness, spasticity and a positive Babinski sign.

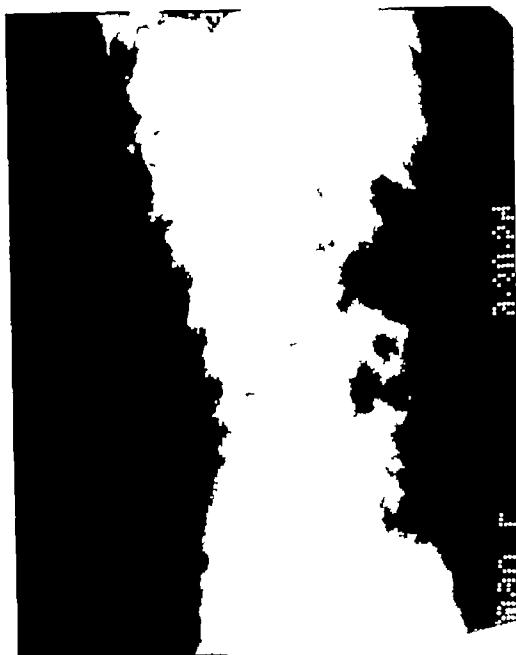


Figure 48 Very early tuberculosis of the spine with thinning of the disc space at the fourth lumbar as the most striking feature. There is also involvement of the vertebral plates abutting the disc

Most authors are agreed that little improvement follows a posterior decompression by laminectomy Conservative treatment alone results in improvement in many cases. Spine fusion adds still others to the list. The operative approach of costo-transversectomy with drainage of the abscess appears most likely to result in prompt improvement of the paraplegia. The ability of the cord to recover from pressure of this type is remarkable with cases recorded of paraplegia of over two years standing with dramatic improvement or complete cure.



Figure 149 Soft tissue abscess causing a paraplegia (*Reprinted with permission from* Gareeau S J and Brady T A. Pott's paraplegia. J Bone & Joint Surg., 32A: 87 1950)

Treatment

One must be quite certain of the diagnosis and those cases which are regarded as possibly having another etiology should be constantly re-appraised in the light of that fact

Needle biopsy is not felt to be indicated. However Johnson brings out the importance of direct surgical biopsy wherever possible as a considerable percentage of cases may be misdiagnosed without it

The patient is suffering from a systemic as well as a local disease The ideal situation for treatment includes institutionalization where bed rest can be enforced and the patient's progress readily reevaluated. Immobilization in hyperextension is not advocated as it unnecessarily prolongs the healing process of a



Figure 160 Spine fusion for tuberculosis of the third and fourth lumbar

disease which is attempting to heal by bony union. Various types of frames holding the patient in a more normal position but one which does not aggravate the deformity have been used the simplest type is an anterior and posterior plaster shell which permits nursing care but aids immobilization The patient may be turned from front to back in these shells

A high calorie high vitamin diet is instituted. Good nursing care is an important adjunct Sunshine and fresh air though aids to good health do not occupy the center of the stage of treatment as formerly

Chemotherapy has now become an important adjunct. The use of streptomycin para amino salicylic acid and iso nicotinic acid hydrazide have been discussed elsewhere Other antibiotics helpful in tuberculosis are already on the

scene It has not been possible to supplant surgical measures with them, but under their cover the approach has become bolder

A safe course to pursue means observation of the child on this regime until definite progress in the direction of healing is obtained. This includes elimination of fever weight gain, declination of the sedimentation rate and a normal lymphocyte-monocyte ratio At this juncture it is felt that the course can be influenced by fusion of the involved vertebra.

Since it has been shown that the disease frequently involves adjacent vertebra that exhibit no pathology by roentgenogram a safe rule is the addition of two vertebrae above and below the lesion to the fusion. Such a rule can be varied depending on anatomic location, deformity, activity of the disease *etc.* Fusion posteriorly is usually followed by fusion anteriorly and eventual cure of the disease After the fusion the patient must still remain on the general measures instituted until the disease is healed. Fusion by itself will not substitute for the general treatment of tuberculosis

Fusion of the spine for tuberculosis was instituted by Hibbs in 1911 By Hibbs method multiple small chips were raised from the laminae, the articulations were curetted and denuded and the spinous processes split and bent over the interspinous area. Albee also reported a spine fusion operation the same year in which an autogenous cortical tibial graft was inserted between the split spinous processes. There have been many variations described since, but fusion based on Hibbs' original concept has proven more successful.

To a Hibbs' type fusion an osteo-periosteal shaving which is continuous has been added with benefit. This is particularly true where the area of attempted fusion involves considerable deformity, the apex of which needs much support to fuse

Sinus formation has proven responsive to chemotherapy particularly those with secondary infections in addition to the tuberculosis. In general abscesses are watched conservatively However large abscesses pointing so that fluctuation is readily elicited may be drained with an attempt made to collapse the walls by compression Streptomycin is introduced into the cavity at the time of drainage Abscesses requiring drainage are usually from lumbar rather than dorsal spine tuberculosis

References

Albee, F H. Transplantation of a portion of the tibia into the spine for Pott's Disease. J A M A., 57: 885 1911

Bosworth D M Della Pietra, A and Rahilly G Paraplegia resulting from tuberculosis of the spine J Bone & Joint Surg., 35A: 735 1953

Garceau G J and Brady T A. Pott's paraplegia. J Bone & Joint Surg., 32A: 87 1950

Girdlestone, S R. The operative treatment of Pott's paraplegia. Brit. J Surg., 19: 121 1931

Hibbs, R. A An operation for progressive spinal deformities. New York State J Med., 93: 1013 1911

Johnson R W., Hillman J W and Southwick W O The importance of direct surgical attack upon lesions of the vertebral bodies, particularly in Pott's disease. J Bone & Joint Surg., 35A: 17 1953

Orell, Svante and Blacklund, Vidar Diagnosis and exact location of lesions by planigraphy in surgical treatment of tuberculosis, spondylitis under cover of streptomycin. Acta chir scandinav 106: 61 1953.

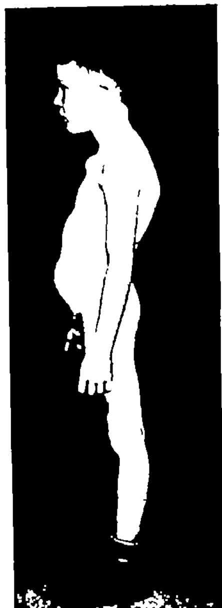


Figure 151 Developing dorsal wedging round back

PRE-ADOLESCENT AND ADOLESCENT KYPHOSIS

Pre-adolescent and adolescent kyphosis is the disability due to dorsal wedging round back that arises in the adult and is both cosmetic and functional. The underlying condition exists in childhood, however Kyphosis juvenilis Scheuermann s disease and vertebral epiphysitis are other names for the condition.

Severe end stages of the condition are recognized in adolescence when the wedging of the vertebra, irregular ossification of the vertebral epiphyses and irregularity of the superior and inferior cortical plates of the vertebra may be striking X ray features

It is particularly helpful to pick out youngsters below the age of twelve who are developing round back deformity so that preventive measures can prevent further deformity Persistent anterior vascular grooves recognizable by roentgenogram leave the patient susceptible to further wedging.

as dorsal vertebrae is closed in over sixty per cent of the children reaching six years of age. The wedging that develops in the fifth, sixth and seventh dorsal vertebrae to form a so-called normal kyphos results in closure of the grooves in the upper dorsal area.

By the time the age of ten was reached only seventeen per cent have some vestige of a groove remaining. Seventy-one per cent have developed a round back in the fifth, sixth and seventh dorsal vertebrae area The vestigal grooves are below this area in the dorsal spine. The lumbar spine grooves have long since been closed

The indentation in the anterior border of the vertebra seen by the roentgenogram has been found by dissection to be occupied by a large endothelium-lined vascular lake formed by the confluence of veins at this point. There is some cartilage also present in the younger children. If the soft tissues are removed from the vertebral body, the groove depth will vary, depending on the degree of closure

Clinical Picture

The patient is often first seen as a posture problem. There may be a tired, aching sensation in the region of the dorsal spine. The round back due to accentuated dorsal kyphos is noted. There may be tenderness of the spine in this area. Accentuated forward bend may be painful Tenderness may be localized to the region of the interspinous ligaments. The hamstrings may be tight and limit straight leg raising



Figure 154 Dorsal spine flexion on forward bend with tight hamstrings. (*Reprinted with permission from* Pediatric Clinics of North America W B Saunders Co., Philadelphia, fall, 1955)

Roentgen Picture

Below ten and before the appearance of the epiphyses of the vertebra there may be wedging of vertebrae below the fifth to seventh dorsal vertebrae. Such wedging added to the wedging above produces the accentuated round back. The anterior vascular grooves may be persistent or closed leaving a linear area of increased density in their wake. Over ten the superior and inferior plates of the involved vertebrae may appear irregular. The epiphyseal plates tend to ossify irregularly in the affected area of wedging.

Treatment

The child who appears to be developing an excessive lumbar lordosis and an accentuated dorsal round back posture and who is then found on a lateral film of the dorsal spine to have wide open anterior vascular grooves becomes the object of special consideration.

There are preventive measures to relieve dorsal spine stress. Sleeping posture is important and any positions which flex the dorsal spine should be avoided. Bed boards and sleeping without a pillow help maintain a flat surface. Relaxation flat on the back on the floor for twenty minute periods relieves dorsal spine stress when tired. Exercises to tilt the pelvis obliterating lumbar lordosis and stretching of tight hamstrings are helpful.

As a constant means of relieving stress in the upright posture we have used a low back brace which flattens the lumbar spine thus resulting in a tendency to extend the dorsal spine as the patient looks forward. In the case of the pre-adolescent this is used until the anterior vascular grooves are closed. In the adolescent it is used until the epiphyses are joined to the parent vertebra and the symptoms relieved.

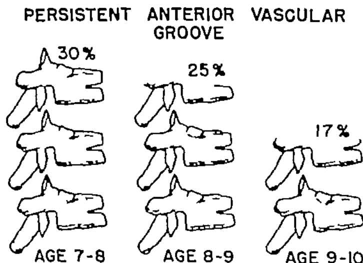


Figure 155 Diagram of decreasing incidence of anterior vascular grooves in the dorsal spine (*Reprinted with permission from* Ferguson A. B Jr The etiology of pre-adolescent kyphosis. Dorsal wedging round back in pre-adolescents. J Bone & Joint Surg., 38A: 149–157 Jan 1956.)

The use of plaster body jackets and even spinal fusion is necessary in some cases to relieve the symptoms.

SCOLIOSIS

WILLIAM F DONALDSON Jr., M.D

The literature concerning scoliosis is voluminous It is filled with much that has been based upon empiric thought both as to etiology and treatment. The wide variety in the methods of conservative therapy found in the literature is in itself a monument to the fact that no one form of therapy has proved to be effective The number and types of braces that have been developed for the treatment of scoliosis are too numerous to mention. We all owe much to such men as Cobb Ferguson, Hibbs, and Risser for their work in this field. We are deeply indebted to Ferguson and Cobb for the development of their systems of measurement of the curves in scoliosis. Without such a system, the accurate follow up of a patient is difficult and inaccurate, and the planning of surgical therapy becomes empiric and almost unscientific.

We all recognize our debt to Hibbs for his development of a technique of spine fusion and its application to the surgical therapy of scoliosis To Ferguson Hibbs, Risser, and their co-workers we owe our thanks for the development of the most efficient method for correction of a scoliosis the turn buckle jacket. Finally we are especially indebted to Cobb for his work in scoliosis and his documentation of the natural course of the disease.

Scoliosis has been defined as any lateral curve tilt or angular deviation of one or more vertebral segments of the spine from the normal straight position. Scoliosis may be divided into two major and distinct groups functional and structural

A functional curve is one that can be completely corrected by the patient *and this correction maintained voluntarily in the erect position. In this type* of scoliosis there is no intrinsic involvement of the mechanics of the spine. There is no structural abnormality of bone nerve or muscle elements of the spine Conversely in structural scoliosis there is intrinsic involvement of bone, and/or muscle and/or nerve elements that support the spine. In a structural scoliosis complete correction cannot be obtained and maintained by the patient in the erect position.

FUNCTIONAL SCOLIOSIS

Functional scoliosis may be seen in a variety of conditions. It may be purely poor posture or it may represent a compensatory adjustment for an organic angular deformity such as a short leg a fixed pelvic tilt secondary to abduction or adduction contracture of the hip "sciatic scoliosis", and so on. These functional curves are not precursors to the development of structural curves Their nature must be understood and their significance appreciated if one is to evaluate properly the patient with scoliosis. (Fig. 1)

By x ray a functional scoliosis has different characteristics from that of a structural scoliosis. For example, there is not seen in a functional curve any evidence of wedging or other structural elements. No treatment is indicated for the functional scoliosis itself



Figure 156 Diagrammatic illustration, functional scoliosis secondary to a short right leg with resultant pelvic tilt.

STRUCTURAL SCOLIOSIS

Etiology

The classification of structural scoliosis according to Cobb has in our hands been an extremely valuable and workable outline

1 Osteopathic
   A Congenital
   B Thoracogenic (postempyema and postthoracoplasty)
   C Other osteopathic
2 Neuropathic
   A Congenital
   B Postpoliomyelitis
   C Neurofibromatosis, syringomelia, etc
3 Myopathic
   A Congenital
   B Muscular dystrophy
   C Other myopathic
4 Idiopathic

The importance of the correct etiologic diagnosis cannot be overemphasized. It must be remembered that scoliosis is only a physical finding and is not in itself a diagnosis.

The etiology of the scoliosis has much to do with its characteristics, and prognosis The fact that the largest segment of the group must be labeled idiopathic does not detract from its value but rather emphasizes the importance of recog

ruling the others The classification is based upon the systems that may be involved etiologically in the development of the scoliosis. The three systems are the bones, muscles and nerves. Subdivisions under each of these three include first the congenital abnormalities and second, the most acquired common etiology For example, under neuropathic, poliomyelitis is by far the most common acquired etiologic neurologic factor Following this are listed the less commonly involved disease, but in most instances the recognition of the coexistence of these entities is extremely important in the management of the whole patient and in particular in the management of the scoliosis. We must always remember that in the treatment of scoliosis we are not dealing with an x ray filled with angular deformities but rather with a patient who has as one thing the physical finding of scoliosis In the past many men have proposed a wide variety of etiologies for idiopathic scoliosis We are all familiar with the textbook picture of the newspaper boy with his heavy load of papers carried over his shoulder This illustration always appears beside a photograph of an x ray showing a scoliosis which the author relates to this supposed etiology To ascribe this as cause and effect is to ignore the mechanics of the spine as related to scoliosis It would be mechanically impossible to demonstrate the forces from such a suggested etiology that could possibly produce a three or four pattern curve.

Some have said that the idiopathic scoliosis represents unrecognized post poliomyelitis scoliosis One can readily demonstrate that this is not true on numerical basis. Idiopathic scoliosis occurs predominately in the female(approximately eighty five percent are female) while known postpoliomyelitis scoliosis occurs about fifty percent male and female. Idiopathic scoliosis occurs within definite age limits, while postpoliomyelitis scoliosis may develop at almost any age The spine in idiopathic scoliosis is almost always stable while a large percentage of the postpoliomyelitis scolioses are unstable and demonstrate marked telescoping and collapse of the spine in the erect position The characteristic curves of these two scolioses are different, and the prognosis and behavior of the curve in each instance is different. For example postpoliomyelitis scoliosis may increase after growth is completed or it may originate after growth is completed. Idiopathic scoliosis, on the other hand, may increase only before the completion of growth and apparently does not have its onset afterwards

Still others have postulated that nutritional lack is the responsible factor This has never been proved by extensive biochemical studies. It is difficult to prove either a nutritional or endocrine variation. In a study of a large segment of patients with scoliosis at any one time the vast majority will show no increase in their angular deformity while under observation. Therefore what variations that might occur would be relativly infrequent and difficult to evaluate and check.

Much has been written concerning the transformation of a functional scoliosis into a structural. In spite of all the papers written with this postulate none have ever shown a true transition from one to the other Even the earliest x rays available in true structural scoliosis will usually show structural changes.

History

In evaluating the patient we are interested in having a complete and accurate history not only as it pertains to the scoliosis but also to the general condition of the patient.

CHART 1
Scoliosis History

Name Date History of Number

Date of Birth Age Sex

COMPLAINT (High shoulder prominent shoulder blade high or prominent hip prominent chest or breast short leg curvature poor posture awkward walk pain in back general fatigue etc)

PRESENT HISTORY Which deformity noted first
Age first noted by whom (patient parents doctor teacher nurse)
Age first treated what treatment (corset brace frame exercises traction)
Duration of treatment
Progress of deformity (stationary increasing decreasing rapid slow)

FAMILY HISTORY Give history of scoliosis or other deformity in each case (other side)
Maternal grandmother (descent) Paternal grandmother (descent)
grandfather (descent) grandfather (descent)
Father Age standing height sitting height
Mother Age
Brothers 1 Age
(names) 2
3
4
Sisters 1
(names) 2
3
4

PAST HISTORY Birth difficult or normal Evidence of trauma?
General health (robust weak normal weight, underweight overweight good color pale sickly frequent illness) Illness—(age and duration) scarlet fever measles mumps chicken pox whooping cough , polio rickets diphtheria empyema pneumonia torticollis Friedreich's ataxia chorea tonsillitis menses age onset (regular irregular scanty profuse duration)

EXAMINATION Date
General posture (good, fair poor) General development (good fair, poor) Musculature good, fair poor) Nutrition (good fair poor) High shoulder (rt left) Prominent scapula (rt left) Prominent low ribs (rt left) Exaggerated flank crease (rt ,left) Prominent hip (rt left) High hip (rt left) List (rt left) R A L A. Rotation (no mild moderate severe)
Standing height Sitting height Weight
Flexibility of spine (flexible moderately fixed fixed)
Curve (Note muscle imbalance on other side of sheet)
Curve corrected on forward bending? (No slight moderate, complete)
Curve corrected on suspension? (No slight moderate complete)

DIAGNOSIS—Scoliosis (check which)
- A Functional
- B Structural
  - I Osteopathic
    - 1 Congenital
    - 2 Thoracogenic
    - 3 Other osteopathic
  - II Neuropathic
    - 1 Congenital
    - 2 Post polio
    - 3 Other neuropathic
  - III. Myopathic
    - 1 Congenital
    - 2 Muscular dystrophy
    - 3 Other myopathic
  - IV Idiopathic

Physical Findings

The physical examination of the patient must again not be limited to the scoliosis, but must be a complete and general examination If this is not done, the etiological background of those cases with known etiologies will be missed, and therefore, the proper management of the patient not determined.

X-rays

The original x ray studies should include an antero-posterior view of the spine, standing, sitting, and recumbent. These should be made on large films at least fourteen inches by seventeen inches in order that the entire curve pattern, as it exists from D 1 to sacrum, may be visualized. Segmental films do not permit

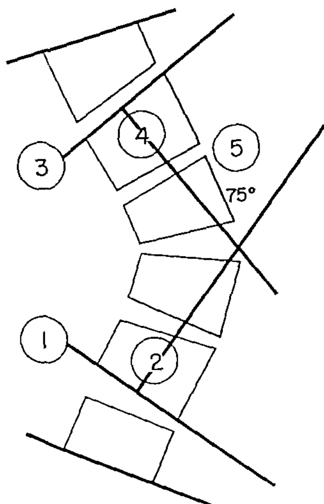


Figure 157 Scoliosis, Measurement of Curve. (*Courtesy of* J R. Cobb M.D.) 1 Bottom vertebra Lowest one whose bottom tilts to concavity of curve 2 Erect perpendicular from bottom of bottom vertebra. 3 Top vertebra Highest one whose top tilts to concavity of curve 4 Drop perpendicular from top of top vertebra. 5 Measure intersecting angle.

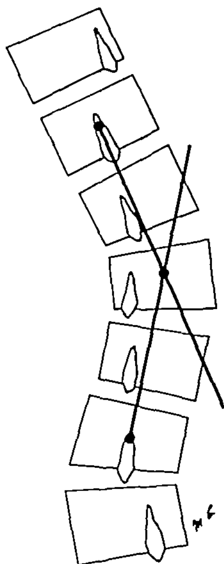


Figure 168 Scoliosis Measurement of Curve (*Courtesy of* A B Ferguson Sr., M D) The central point of each end vertebra and of the apical vertebra is marked The end vertebra are defined as the first non rotated vertebra identified by the position of the spinous processes. The lines from the central point of the apical vertebra to that of each end vertebra form an angle The deviation from these lines from 180 degrees is the angle of the curve

accurate measurement of the curves. A lateral of the spine must also be made as part of the original x ray study Follow up films need only include antero-posterior standing and sitting x rays of the spine and should be made at three-month intervals The erect films of the standing and sitting position will serve as a counter-check one against the other By either of the methods of measurement (figs 167 and 168) the variation between these two films usually is not more than five degrees except in the lumbar area where the spine is more flexible

All the curves must be measured and these findings should be recorded at each time for comparative study in the future Additional films such as sitting tilt and lateral bending films are chiefly of value in the preoperative evaluation of the patient, prior to correction and fusion if this procedure is indicated or if the patient appears to be losing flexibility in the opposite curves to the major curve.

CHART 2

Scoliosis record

NAME

DIAGNOSIS

DOCTOR

HISTORY NO

Date Age Ht Standing Ht Sitting Weight L A R. A. List High Shoulder Low Hip Prominent Hip							
X ray Date Born Age							
( ) Curve {Standing Sitting Supine} Rotation (+ ++ etc) Wedging (+ ++ etc)							
( ) Curve {Standing Sitting Supine} Rotation (+ ++ etc.) Wedging (+ ++ etc)							
( ) Curve {Standing Sitting Supine} Rotation (+ ++ etc) Wedging (+ ++ etc)							
( ) Curve {Standing Sitting Supine} Rotation (+ ++ etc) Wedging (+ ++ etc)							

Procedure

Idiopathic Scoliosis

In a large scoliosis clinic where all scolioses are seen and followed, not just the severe ones, the vast percentage of the cases will be idiopathic in nature. Idiopathic scoliosis is a self limited disease characterized by the development of a structural scoliosis for which there is no known etiology. The angular deformity does not progress at a constant rate, but rather at unpredictable intervals. If an increase does occur it almost always occurs before age fifteen in females or sixteen in males. Progression may or may not occur, and if it does it may stop spontaneously with or without treatment. Growth in itself is not the only factor involved in the increase in the angular deformity of an idiopathic scoliosis. Many patients will show a marked increase in height without any significant change in the degree of angular deformity.

If a curve is going to increase its angular deformity, it will do so while growth is occurring but not because of it. These two, then, are concomitant occurrences and not cause and effect.

The observation by Risser that after the iliac crests are fully capped by their apophyses idiopathic scoliosis does not increase is of particular value in those patients in whom the spine matures before the age of fifteen or sixteen.

Approximately eighty five percent of the cases of idiopathic scoliosis occur in the female. Many of these have a very early or very late onset of sexual maturity as evidenced by the menarche. Many of the males who develop idiopathic scoliosis have a fairly feminine type of build, at least during the active growth period.

There is no accurate evidence available that any system of exercises, braces, or casts will stop a curve from progressing if it going to do so. We cannot take credit for those that do not increase, because idiopathic scoliosis is characterized by intermittent periods of increase. Nor can we take credit for those whose idiopathic scoliosis stops spontaneously. There is no well documented evidence that significant correction by any conservative means has ever been maintained.

Until a large series of cases of true idiopathic scoliosis can be presented, proving that the incidence of increase in comparative cases is less in those wearing a brace or other form of immobilization than those that do not, we cannot claim that the conservative therapy has much to offer the patient with idiopathic scoliosis.

Intelligent observation is probably the most valuable thing we have to offer these patients. Rest in recumbency seems to have some beneficial effect.

The operative treatment of a patient with idiopathic scoliosis must be based upon an accurate knowledge of the mechanics of the spine, both the normal and the abnormal, as related to scoliosis. The selection of patients with idiopathic scoliosis for correction and fusion requires a thorough understanding of all the problems involved.

This procedure is not indicated in idiopathic scoliosis as a preventive measure in a patient with a mild curve in whom serial x rays show that there has been no increase in the angular deformity. It is indicated in those in whom the angular deformity when first seen is severe enough to make it desirable or in whom there has been a rapid and significant increase. The procedure may be contra indicated because the amount of correction obtainable is not significant, for example twenty degrees or less or where no correction of the major curve is permissible. It may also be contra indicated by the presence of other organic or psychological conditions that make any surgical procedure unwise.

The ideal age for performing this procedure is between the ages of twelve and sixteen. However, as we must not fuse unnecessarily in the mild nonprogressive case we must also not allow the patient to become progressively deformed to the point of no longer being able to salvage the patient cosmetically and physiologically before undertaking the procedure (fig. 159).

The modified Risser type of turn buckle jacket has been the most satisfactory means developed to date for the correction of a structural scoliosis (fig. 160).



Figure 159 Left M. W. 8-year-old female, idiopathic scoliosis 1948. Treatment consisted of the use of exercises and braces. Operative intervention never suggested. *Right* x-ray of same patient taken in 1953, patient now age thirteen. Correction and fusion resulted in very definite improvement but certainly the ideal time for this procedure was missed in the interim. Serial x-ray observation at three month intervals would have graphically demonstrated the progression of angular deformity and indicated the need for surgical therapy.

The preoperative evaluation of the patient must include not only a complete history and physical and x ray studies, but also certain baseline laboratory studies for the preoperative evaluation of the patient and for the postoperative regulation of the patient who may in the postoperative period, develop any disease We routinely obtain a complete blood count with differential the urinalysis, sedimentation rate serology for Luetic Infection, total protein and Ag ratio, electrocardiogram, chest x ray, and vital capacity

The amount of correction permissible may be determined by the use of standing and recumbent bending films or sitting tilt films (fig. 162)

After it has been determined that the patient exhibits the required criteria for correction and fusion, the patient is placed in a Risser turn buckle jacket (fig. 160) Care must be taken in the application of the jacket that all areas are well padded and that the hinges are placed eccentrically to the apex of the curve to be corrected The more acute the angulation, the more eccentrically the hinges should be placed Following the application of the cast the patient must be followed closely in order to avoid unnecessary complications, such as pressure sores The particular areas in which these may develop are anterior superior spines of the ilium, the prominent ribs on the convexity, and the chin

Correction must be obtained in a slow and gradual manner Correction should be painless X rays should be made through the cast at intervals to determine the amount of correction obtained until the maximum amount of correction that is permissible or obtainable has been reached

The jacket should be filled in. The window is then cut out in the posterior aspect of the cast over the operative area.

The spine is marked at two levels at either end of the fusion area. We have found the methylene blue marker, and metal marker technique to be most satisfactory

The selection of a fusion area in an idiopathic scoliosis must be done with extreme care if we are to have satisfactory results. Where a complete permissible correction has been obtained, the fusion area in a stable idiopathic scoliosis is usually the major curve only If it has not been possible to obtain the maximum amount of correction permissible then the inclusion of some compensation by the addition to the fusion area of an extra segment or segments from the adjacent opposite curve or curves may be desirable. (fig. 165 to 171)

The operative procedure, spine fusion is carried out under endotracheal anesthesia usually administered through a intranasal endotracheal tube Usually 1500 cc of typed and cross-matched blood is available The blood loss is followed by the technique of weighing the sponges The operative area is prepared and draped through the window in the jacket. The previously identified and marked posterior vertebral spines are surgically marked (fig. 169) A modified Hibbs type of fusion is performed We have used cancellous bone bank bone as supplementary bone to provide a larger and more massive fusion mass.



Figure 160 Upper modified Risser turn-buckle jacket with full head piece and thigh extension on the side of the convexity of the major curve The anterior and posterior hinges and the position of the turn-buckle can be seen *Lower* a modified Risser turn-buckle jacket viewed from the side of the convexity of the major curve Note the eccentric position of the anterior and posterior hinges. These hinges are made of aluminum and are therefore relatively radiolucent.

When one considers that it is the mature fusion mass that must be counted upon to maintain the corrected position against all the forces that have produced the curve the necessity for a mature massive fusion is apparent. The operative procedure can usually be performed in one or possibly two stages

Postoperatively the patient is immobilized in the jacket for approximately six months During this time the patient remains recumbent. At the end of six months the cast is removed and x rays are made to determine the maturity and continuity of the fusion mass. A regular body jacket is then applied and the patient is allowed up or required to remain recumbent, depending upon the

maturity and the continuity of the fusion mass as demonstrated by x ray If there is any doubt as to the solidity of the fusion, the patient remains recumbent in the body cast for another two to three months Comparative x rays are made at that time and the patient is allowed up if the fusion mass is mature enough The body jacket may usually be discarded in a few months. It has been our experience that, with a solid mature fusion, there is no appreciable loss of correction.

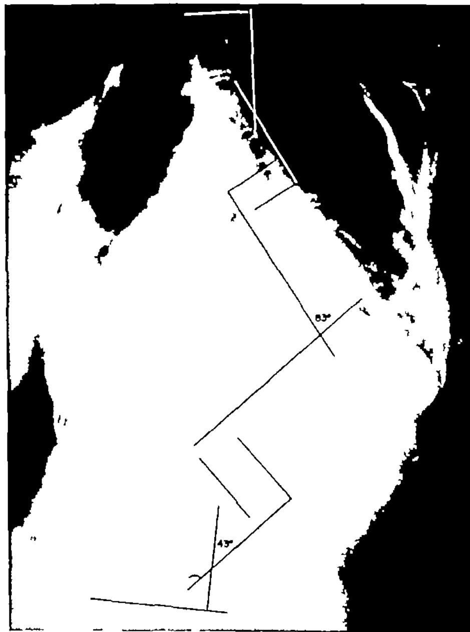


Figure 161 S T age 14 with a typical idiopathic scoliosis. Major curve D6 to L2, 83 degrees. Minor curves, D1 to D6 29 degrees L2 to S1 43 degrees.



Figure 162 Same case as in Figure 161 Recumbent left bending film Minor curve D1 to D6 reduces to 16 degrees L2 to S1 reduces to 10 degrees. This demonstrates considerable flexibility in the opposite curves and indicates that considerable reduction in the angular deformity of the major curve D6 to L2 is permissible.



Figure 163 Same case as in Figure 161 Postoperative film. Major curve D6 to L2, 30 degrees. This degree of correction has been maintained.



Figure 104 M K., age 13 typical idiopathic scoliosis

Figure 105 Continuation of case in Figure 164 Preoperative film Major curve D6 to L1 89 degrees. Minor D1 to D6 53 degrees L1 to S1 36 degrees

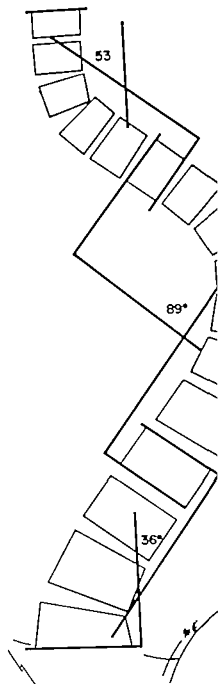


Figure 106 Diagrammatic illus
Figure 165



Figure 167 Continuation of case in Figure 164 Preoperative lateral recumbent bending film Minor D1 to D6 30 degrees L1 to S1 1 degree.

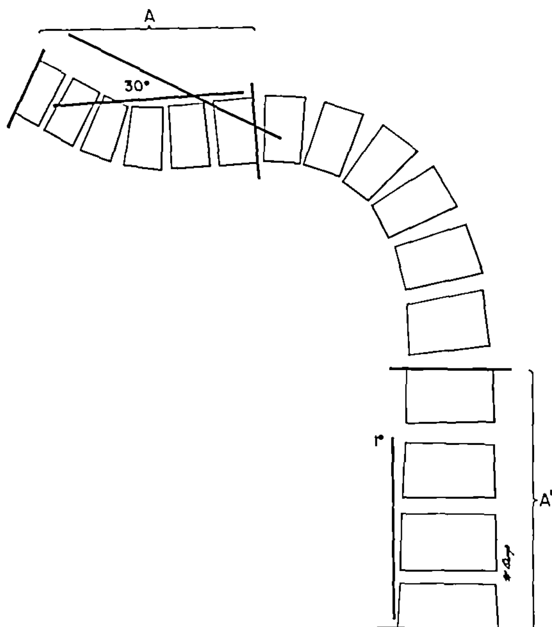


Figure 168 Diagrammatic illustration of Figure 166



Figure 169 Continuation of case in Figure 164 Preoperative x-ray showing reduction in the major curve D6 to L1 from 80 degrees to 34 degrees. Note the metal markers over the spinous processes of D6 and L1 The film is made through the Risser jacket. The opaque discs are the studs in the hinges. The central opaque disc is at the apex of the anterior hinge, the posterior hinge having been removed in excising the window for surgery It is to be noted that even when the desired correction has been obtained, the apex of the hinge has just reached the concavity of the major curve from its original position eccentric to the convexity and at or above the apex of the major curve

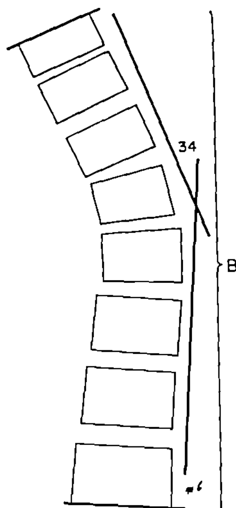


Figure 170 Diagrammatic illustration of Figure 167



Figure 1 1 Continuation of case in Figure 164. Composite film showing the eventual angular pattern of the patient on completion of correction and fusion. The top and bottom portions, the minor curves are taken from Figure 166 bending film Superimposed between these is our preoperative correction film, Figure 167 By this means we can determine that our patient is not over corrected. More correction would have been permissible if it had been obtainable Serial x-rays taken during correction showed that no further correction was obtainable

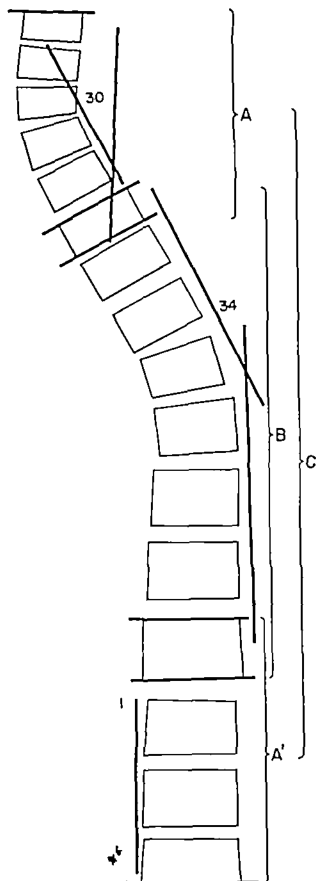


Figure 178 Diagrammatic illustration of Figure 168

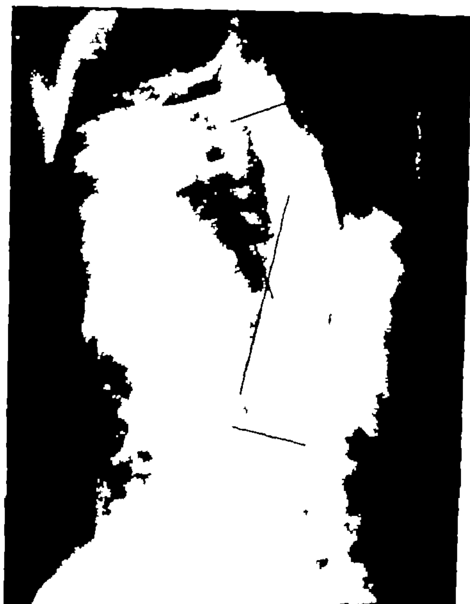


Figure 173 Continuation of case in Figure 164 Postoperative films demonstrate maintenance of correction obtained. Note the neurologic clips which have been placed on the remaining tips of the spinous processes at the ends of the fusion area D6 to L1 These readily identify the area fused in the follow-up x-ray studies.

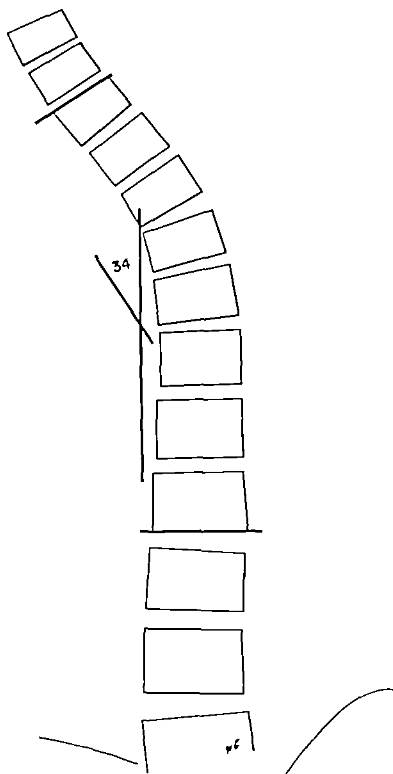


Figure 174 Diagrammatic illustration of Figure 169



Figure 165 Continuation of case in Figure 164. Oblique films demonstrating the fusion mass, D6 to L1 Again note the neurologic clips.

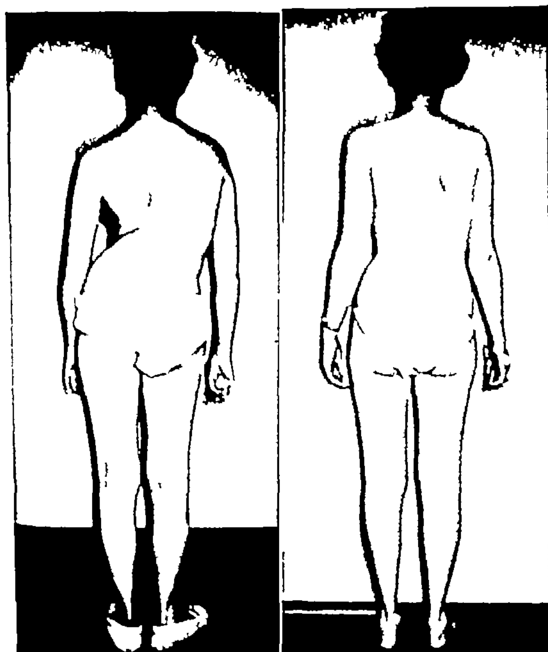


Figure 176 M K. PA photographs. Left preoperative. Right postoperative

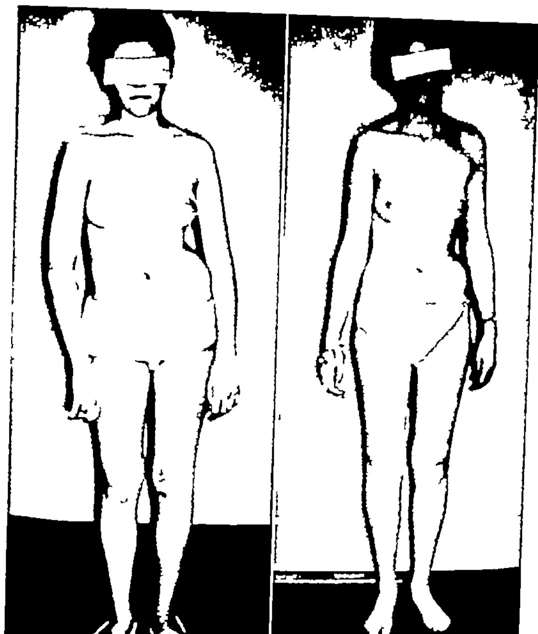


Figure 177 M K. AP photographs. Left preoperative Right postoperative

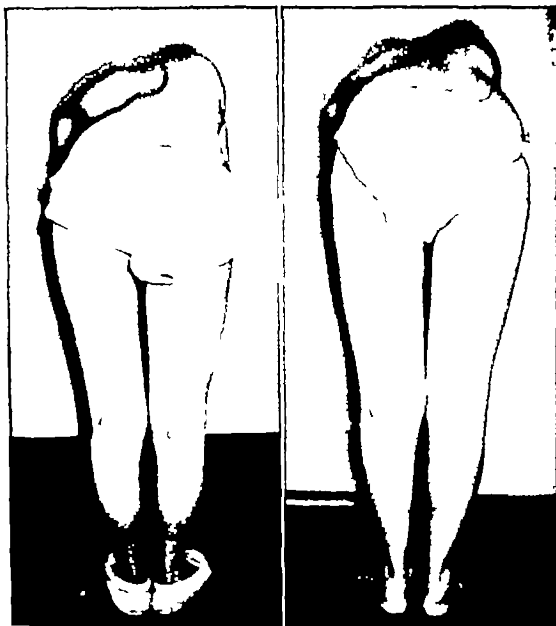


Figure 1 S M K Left preoperative forward flexion photograph showing the amount of rotational deformity in the dorsal and lumbar regions Right postoperative photograph showing some decrease in the rotational deformity of the dorsal and lumbar regions

Postpoliomyelitis Scoliosis

The cases of postpoliomyelitis scoliosis account for approximately ten per cent of those seen. Many of these will have unstable spines and, in these correction and fusion may be indicated for stability only.

All patients who have had poliomyelitis must be watched for the development of scoliosis. Some of these do not develop a clinically evident scoliosis for as long as ten years after the onset of their poliomyelitis. One cannot predict which patients will develop scoliosis by the muscles involved. The muscles attached to the spine are numerous and their possible combination of involvement and their effect upon the spine cannot be comprehended. The use of bracing and casts in the postpoliomyelitis scoliosis may have definite value in providing stability for the trunk. They will not stop the curve from progressing if it is going to do so.

In selecting the fusion area in these patients at least the major curve and a segment or more below must be included. At times it may be necessary to fuse even further in order to provide the stability necessary. In some patients, it may be desirable to fuse to the sacrum. When this is done it may convert some patients to being good sitters and nonwalkers. When this is undertaken the physician must be sure that this will be definitely desirable for the patient. It is much better to be a good sitter than to be a poor occasional walker and a poor sitter. As in all patients with scoliosis, the individual patient must be individually evaluated and his or her therapy made applicable in view of this evaluation.

Congenital Scoliosis

In evaluating these patients one cannot add up the segmental errors and determine the prognosis. It must be remembered that the x rays show only the bony variations (fig. 180 and 181) and not the associated soft tissue anlagal changes. Most congenital scolioses are stable. The resection of hemivertebrae in the reported series carries a high morbidity. Again we must remember that the evident hemivertebrae is only one part of the problem. If a significant curvature does develop it may well develop in the area of the greater soft tissue anlagal variations. In a series reported by Cobb about thirty per cent of his patients required correction and fusion.

Von Recklinghausen's Neurofibromatosis

The curve pattern in a patient who has a scoliosis associated with a neurofibromatosis is usually characteristic. The curve will present a short, sharp, angular pattern by x ray and the structural changes will usually be excessive to the amount of angular deformity present. This is particularly evident in the milder curves. (fig. 182) Many of these patients will show the cutaneous manifestations of neurofibromatosis, café-au lait spots, subcutaneous neurofibroma or pedunculated neurofibroma. Almost all of these patients will require correction and fusion. It has been the experience of many that these curves usually progress and eventually produce the most severe deformity seen in the patient with scoliosis. Many feel that once the diagnosis is made correction

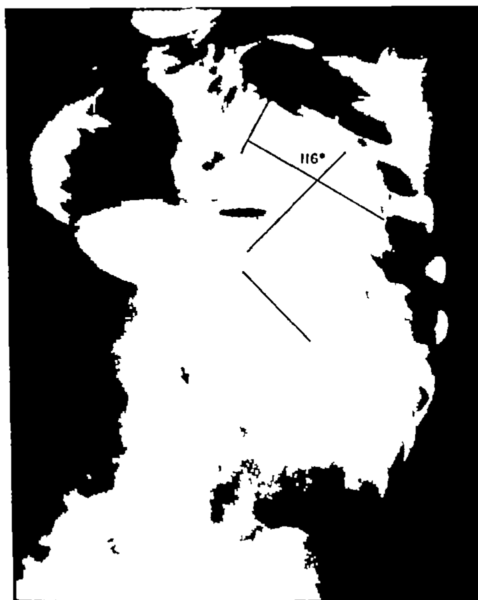


Figure 170 C W 5-year-old female with congenital neuropathic scoliosis Major curve D5 to D11 116 degrees. Serial x-rays have not shown any progression of the angular deformity but repeated neurologic examination revealed development of diminished sensation of L5 S1 and progressive loss of function in the right peroneals On the original examination and for a year thereafter the extremity neurologic examination had been negative



Figure 180 Same case as Figure 179 Myelogram demonstrating diastematomyelia level of L1 Surgical correction resulted in return of sensation and no further progression of peroneal weakness.

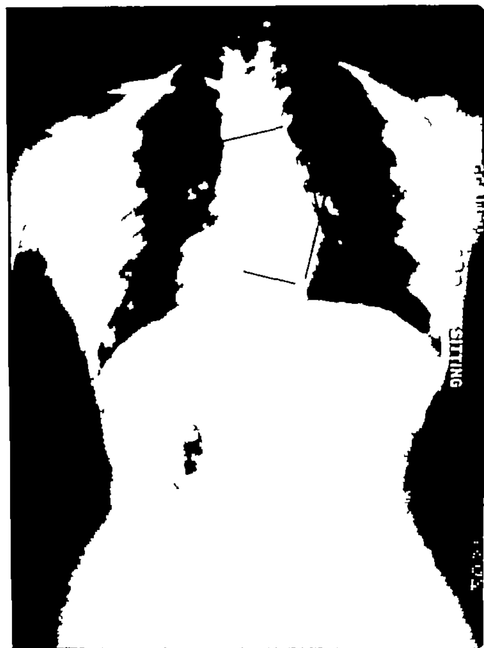


Figure 181 Congenital scoliosis with multiple errors in the segmentation of the thoracic spine Serial films have not shown any progression in the degree of angular deformity



Figure 188 Neuropathic scoliosis—Von Recklinghausen. This patient has skin manifestations of neurofibromatosis. There is a short, fairly sharp angular major curve. The curve has structural changes in excess of the amount of angular deformity. It is fairly typical of mild scoliosis secondary to neurofibromatosis.

and fusion should be performed This stems from the appreciation of the marked deformity that usually develops if this is not done

In the patient with a neurofibromatosis and scoliosis, one must not consider performing a laminectomy for relief of the pressure symptoms of the neurofibroma, if these are present, before performing a spine fusion It has been the experience of those who have performed such a procedure prior to spine fusion that the results are disastrous

Postemphyema and Post Thoracoplasty Scoliosis

Postemphyema and post thoracoplasty scoliosis is becoming less of a problem because of the decreased incidence of these entities. It should always be looked for, and the patient followed with serial films Correction and fusion are indicated in those in whom a significant curve develops or in whom definite progression is demonstrated It is usually in children that significant curves develop

Muscular Dystrophy

The patients with muscular dystrophy who develop scoliosis often develop an extreme amount of deformity The recognition of the associated disease with the scoliosis is extremely important These patients must not be placed in recumbency for the period required for correction and fusion or we will find that we have a patient whose spine is straighter but whose muscular atrophy has progressed to the point that he may no longer be capable of walking.

SUMMARY

In summary, it would seem that the intelligent management of a patient with scoliosis is dependent upon the appreciation of etiologic basis. The vast majority of the patients will not require correction and fusion When correction and fusion are indicated it must be carefully planned and carried out. If the criteria are met successful correction and fusion can be obtained and the results are gratifying

The author wishes to express his appreciation to Dr John R. Cobb for his personal communications and the many hours spent together as teacher and pupil

References

ALBEE, F H AND KUSHNER, A. Albee spine fusion operation in treatment of scoliosis. Surg. Gynec. & Obst., 66: 797 1938.

ARKIN A. M., PACK, H. F., RAMBOHOFF N S AND SIMON N Radiation induced scoliosis. J Bone & Joint Surg., 32A: 401 1950

ARKIN A. M Mechanism of structural changes in scoliosis. J Bone & Joint Surg., 31A: 519 1949

ARKIN A. M AND SIMON N Radiation scoliosis experimental study J Bone & Joint Surg 31A: 296 1950

BARR, J S Turnbuckle brace 3 point pressure brace for correction and treatment of ambulatory care of scoliosis. J Bone & Joint Surg., 18: 760 1936

BISGARD J D Thoracogenic scoliosis influence of thoracic disease and thoracic operations Arch Surg., 29: 417 1934.

BORDEN J N AND NICHLAS J N Experimental scoliosis role of epiphysis. Surg. Gynec. & Obst., 90: 672 1950

COBB J R. Technique after-treatment and results of spine fusion for scoliosis. The Am Academy of Orthopedic Surgeons, Instructional Course Lectures, 9: 65 1952.

COBB J R. Outline for study of scoliosis. The Am Academy of Orthopedic Surgeons, Instructional Course Lectures, 5 1948.

COBB JOHN R. Treatment of scoliosis. Connecticut M J 7: 467 1943

CAMPBELL, E. L. Excision of hemivertebrae for correction of congenital scoliosis. J Bone & Joint Surg 14: 555 1932

FERGUSON A B Study and treatment of scoliosis. South. M J 23: 116 1930

FERGUSON A B Roentgen interpretations and decisions in scoliosis. The Am. Academy of Orthopedic Surgeons, Instructional Course Lectures 7: 160 1950.

FERGUSON A B Roentgen diagnosis in extremities and spine. The Am Academy of Orthopedic Surgeons, Instructional Course Lectures, 2: 214, 1947

GHORMLEY J N Congenital scoliosis due to unusual deformity of vertebral column. New York State J Med 38: 432 1938.

GUDD F B Scoliosis accompanying chronic infected open pneumothorax its causation and correction. Arch Surg., 5: 366 1922.

HEYMAN C H Spinal cord compression associated with scoliosis. J Bone & Joint Surg., 19: 1081 1937

HIBBS, R. A RISSER, J D AND FERGUSON A. B. Scoliosis treated by fusion operation, end-result study of 360 cases. J Bone & Joint Surg., 13: 91 1931

JAMES J Idiopathic scoliosis. J Bone & Joint Surg 1954.

JAMES, J Two curve patterns in idiopathic structural scoliosis. J Bone & Joint Surg., 1951

KENDRICK, J Correction of scoliosis by use of modified turnbuckle jacket. Cleveland Clin. Quart. 4: 16 1937

KERGIN F G AND DEWAR, F P Pleural decortication in prevention and treatment of thoracogenic scoliosis (resulting from fibrothorax) Arch. Surg., 61: 705 1950.

KLEIN A Treatment of structural scoliosis at Massachusetts General Hospital. J A. M A., 78: 418, 1922

KLEINBERG S A survey of structural scoliosis the principles of treatment and their application. The Am. Academy of Orthopedic Surgeons, Instructional Course Lectures, 7: 130 1950

KLEINBERG, S Structural scoliosis secondary to syringomyelia. J Bone & Joint Surg. 15: 779 1933

KLEINBERG S Paralytic scoliosis analysis of 51 cases. Am J Surg., 64: 301 1944.

KLEINBERG, S Results of spine fusion for scoliosis. *J Bone & Joint Surg., 11: 66*, 1929

KUHNS J G Congenital scoliosis review of 77 patients. New England J of Med., 210: 1310 1934

LAWMAN C L Relation of abdominal muscles to paralytic scoliosis. J Bone & Joint Surg 14: 763 1932

McKENZIE, K G AND DEWAR, F P Scoliosis with paraplegia. J Bone & Joint Surg., 31: 162, (May) 1949

MAYER, L Treatment of congenital scoliosis due to hemivertebra. J Bone & Joint Surg., 17: 671 1935

NACHLAS J N AND BORDEN J N The cure of experimental scoliosis by direct growth control. J of Bone & Joint Surgery 1951

PEABODY C N Correction supports in scoliosis. J Bone & Joint Surg 13: 177 1931.

PEASE, C N Modification of turnbuckle joint for treatment of scoliosis suggestion as to management. J A M A., 86: 1232 1926

PONSETI, J AND FRIEDMAN B Prognosis in idiopathic scoliosis. J Bone & Joint Surg., 32A: 381 1950

RISSER, J C Scoliosis treated by fusion preliminary report on some observations in 343 cases. Am. J Surg., 4: 496 1928.

RISSER, J Important practical facts in treatment of scoliosis. The Am Academy of Orthopedic Surgeons, Instructional Course Lectures, 5: 1948.

RISSER J C AND FERGUSON A B Scoliosis prognosis J Bone & Joint Surg., 18: 667 1936
SMITH A, BUTTE, F L AND FERGUSON A B Treatment of scoliosis by wedging jacket and spine fusion, J Bone & Joint Surg., 20: 825 1938.
STEINDLER A. Nature and course of idiopathic scoliosis The Am Academy of Orthopedic Surgeons, Instructural Course Lectures 7: 150 1950
STEINDLER, A Compensation treatment of scoliosis, J Bone & Joint Surg., 11: 820 1929
ULLRICH H F Operative treatment of scoliosis Am J Surg 45: 323 1939
VAN LACKUM H L AND SMITH A Removal of vertebral bodies in treatment of scoliosis. Surg Gynec & Obst., 57: 250 1933
VAN LACKUM W H The surgical treatment of scoliosis The Am Academy of Orthopedic Surgeon, Instructional Course Lectures 5 1948
VAN LACKUM W H Surgical scoliosis Surgical Clinics of North America. April 1951
WILES P Resection of dorsal vertebrae in congenital scoliosis. J Bone & Joint Surg., 33-A: 151 1951

PYOGENIC INFECTION OF THE SPINE

Pyogenic infection of the spine is rare. It is brought to clinical attention by pain in the back accompanied by muscle spasm. In addition to holding the involved segment of the spine rigidly the patient may exhibit limitation of straight leg raising. There is local tenderness of the involved segment. Past history may reveal a previous lumbar puncture.

The roentgenogram reveals narrowing of the interveretebral disc with areas of destruction along the adjacent vertebral plates. The shadow of an accompanying abscess is not a feature as in tuberculosis. Staphylococci etiology may lead to some bony proliferation. Brucella, paratyphoid, *Bacillus coli* and streptococcus lesions exhibit no bony reaction in the early stages.

Treatment

These cases have responded to immobilization in plaster body and thigh spica with appropriate chemotherapy

Lumbar puncture and diagnostic aspiration is to be avoided in general.

References

COUPERE, E. L., SCHNUTE, W J AND CATTELL, L M The use of penicillin in the treatment of acute hematogenous osteomyelitis in children. Ann. Surg., 122: 954 1945
DICKSON F D The clinical diagnosis, prognosis and treatment of acute hematogenous osteomyelitis. J.A.M.A., 127: 212, 1945
GREEN W T Osteomyelitis in infancy J.A.M.A., 105: 1835 1935

5

The Neck

JOHN DONALDSON M.D

CONGENITAL DEFORMITIES

SPINA BIFIDA

Spina bifida is the term applied to the hiatus or gap due to failure of fusion of the laminae of the neural arches and through which a sac may protrude. This sac may or may not contain nerve elements and the degree of defect determines the type of resulting deformity

Etiology

Spina bifida occulta occurs on occasion in the neck as a congenital skeletal defect without any protrusion of the dura. Occasionally a meningocele originates from the dura and protrudes through the laminal defect perhaps containing spinal fluid but no neural elements.

Clinical Picture

A spina bifida occulta is suspected if superficially on the neck there is a localized overgrowth of hair or a small dimple. Very rarely is there a lump or protrusion sufficiently large enough to palpate.

Treatment

Ordinarily no treatment is indicated.

HEMIVERTEBRA

A hemivertebra is the result of a congenital failure in growth of a half or a portion of the primordium of a normal vertebra.

Etiology

The origin of such deformities cannot be laid to the failure of development of ossification centers, but antedates their formation.

Clinical Picture

The vertebra above and below the hemivertebra compensate for the hemivertebra. Such scolioses ordinarily do not progress with growth. Occasionally,

however, a vertebra can with growth change position because of a missing portion of vertebra below it For this reason braces may be necessary to prevent increase in curvature with growth Such a deformity usually becomes fixed, and increase in curvature does not develop

Treatment

Other than the use of occasional bracing to prevent possible progression, treatment is not as a rule indicated. In some parts of the spine, hemivertebrae have actually been removed but this is not a procedure indicated or suitable in the cervical region

KLIPPEL FEIL SYNDROME

The Klippel Feil syndrome might better be termed congenital fusion in which two or more vertebrae become anatomically fused to each other Such spines appear quite disorganized by roentgenogram.

Etiology

The deformity is a result of failure of normal development of the vertebrae.



Figure 183 Multiple vertebral and rib anomalies in upper thoracic spine in patient with Klippel-Feil syndrome



Figure 184 Short cervical spine in Klippel-Feil syndrome

Clinical Picture

The cervical spine is limited in motion. The neck appears to be short, and if sufficient involvement has occurred, the head may seem to be resting between the shoulders and broad "webs" of skin and soft tissue extend out to the shoulders. There may be some neurologic changes accompanying this deformity due to involvement of the cord X rays reveal vertebral disorganization with multiple anomalies There is no characteristic distribution of the involved vertebrae.

Treatment

Severe though this disability may be, there is no treatment which will fully correct the deformity Stretching by traction and passive exercises are used. Plastic improvement of the skin "webs" may be indicated

SPRENGLES DEFORMITY

Congenital elevation of the scapula is also known as Sprengle's deformity It is characterised by a cephalic location of the scapula as compared to the normal position.

Etiology

This condition appears to be due to a failure of descent of the scapula from its original position in the cervical region, this process occurring during embryonic development. The upper limb bud develops as a mass of undifferentiated mesoderm in the cervical region during the fourth week. Early in the fifth week condensation of scleroblastema of the limb bud has extended to the distal part and the anlages of the scapula, humerus, radius and ulna are distinguishable. In the eleven mm. embryo, the scapula is composed of free cartilage surrounded by a dense blastema and it lies opposite the lower fourth cervical and the upper one or two dorsal vertebrae. In the fourteen mm. embryo the scapula is well developed and is composed mostly of cartilage. It has migrated downward so that less than half of it lies above the first rib. Keith has stated that the scapula begins to descend from the neck to the thorax at the ninth week and reaches its ultimate thoracic position at the beginning of the third month. These facts seem to indicate that the high position of the scapula in persons with this deformity is determined very early at a time when the embryo is so small that it is inconceivable that any external pressure from abnormal position *in utero* or any deficiency of amniotic fluid could have had any bearing.

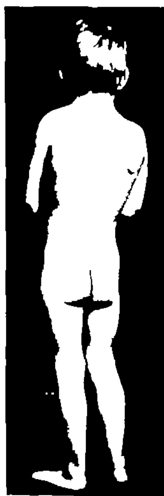


Figure 185. High scapula on the left constituting Sprengel's deformity.



Figure 186 Roentgenogram with congenital high scapula and accompanying low cervical upper dorsal scoliosis.

Clinical Picture

The scapula is shortened vertically and supraspinous portion is hooked forward. In about one-fourth of the cases, a connecting structure is present between the scapula and the cervical portion of the spine. This structure may be fibrous cartilaginous or bony and has been termed the "omovertebral bone. As a result of these vestigial attachments elevation of the arm may be restricted and the whole shoulder seems to be displaced forward. A mild scoliosis of the upper dorsal and low cervical region may also be present.

Treatment

DeForest Smith has emphasized that operative repair is not always advisable and increased range of motion can often be obtained by exercises. If the deformity is not severe, conservative measures are indicated. There have been developed however various procedures which should be considered either to improve a severe deformity or to obtain more motion of the arm.

Operative procedures developed consist of (1) removal of the omovertebral bone alone to improve motion (2) extensive sub-periosteal stripping of the scapula and removal of a large part of the upper portion of the scapula (3) freeing the scapula completely by sub-periosteal dissection and anchoring it to a low level by fastening it to a rib

In this last procedure, some surgeons prefer a one-stage operation and others prefer to correct gradually by traction through a plaster body spica.

The scapula is pulled down into a sleeve of muscles which has been made of



Figure 187 Post operative application of spring traction to inferior angle of scapula. The scapula is freed and pulled down into a surgically created pouch

muscle groups which have been stripped sub-periosteally A temporary partial brachial paralysis may complicate this procedure and the patient must be carefully observed to guard against its occurrence In 1952 C J Jeanopolous recommended that no attempt be made to correct the abnormal position, but instead he carried out only a subperiosteal stripping and a resection of the superior medial portion including the spine and supraspinous portion as well as a large part of the body of the scapula. When present the omovertebral bone was removed as well He felt that a gradual improvement in position would spontaneously take place in some cases following this procedure.

Occasionally an osteotomy of the base of the acromion or of the outer third of the clavicle has been performed to help in getting a scapula down to a lower level and some claim better results by excising the outer end of the clavicle.

Osteotomy at the base of the glenoid and sliding the vertebral portion or greater part of the entire scapula down have been reported allowing healing in this position

CONGENITAL ABSENCE OF MUSCLES OR MUSCLE GROUPS

Absence of a muscle or muscle group may result in contracture This would follow if function is not substituted for the absent muscle by other muscles controlling the part.

Bracing and exercises carrying the part through a full range of motion may be indicated until the function of the absent muscle can be substituted for by transplant if necessary Not all absent muscle groups lead to deformity since it would vary with the anatomic location. Thus absence of the palmaris longus is not significant but unilateral absence of the sternocleidal mastoid may lead to deformity

The most frequent muscle group affected is the pectoral with trapezius next in order Although any muscle may be affected proximal or trunk muscles appear more liable to involvement Absence of the sternocleidal mastoid is not infrequent



Figure 188 Congenital absence of the sternocleidal mastoid muscle The area normally occupied by the sternocleidal mastoid muscle is retracted on inspiration and expanded on expiration.



Figure 189 Congenital absence of the pectoralis major with the creation of an abnormal skin fold across the area normally occupied by that muscle.

CONGENITAL HEMI HYPERTROPHY

Mild forms of asymmetry are evident to some degree in all persons The effects of such involvement may be seen in neck and face

Etiology

One must distinguish carefully between congenital hemiatrophy and congenital hemihypertrophy The latter is detectable at birth and originates in some developmental abnormality It is usually associated with other developmental anomalies. Gesell promotes the theory of an imbalance of the normal process of twinning' as a cause Hemiatrophy can be caused by lesions of the central nervous system such as a brain injury It is known too that arteriovenous aneurysms can cause true hypertrophy

Clinical Picture

In a typical case of hemihypertrophy, the hypertrophied side of the body (face thorax arm, leg etc.) may show a purplish mottling due to vascular engorgement on the affected side

Treatment

Treatment consists of trying to obtain as much symmetry as possible during growth. The growth in the extremities can be controlled as indicated by epiphyseal arrest, lengthening and shortening procedures.

FAMILIAL NUCHAL RIGIDITY

This condition is one of rigidity of the cervical spine It is probably the direct antithesis of Ehler Danlos disease which is one of hypermobility of all joints of the body

Etiology

Ehler Danlos disease which is associated with hyperelasticity of the skin and fragility of the skin and blood vessels has been proved to be an inherited, dominant constitutional dyscrasia It is thought that Familial Nuchal Rigidity is likewise inherited by dominant transmission

Clinical Picture

This condition is characterized by an inability to flex the neck because of relatively short ligaments and soft tissues on the posterior or dorsal aspect of the vertebral column. It is sometimes confused with ankylosing arthritis

Treatment

Treatment is unavailing.

ACQUIRED DEFORMITIES

FOETAL POSITION *IN UTERO*

Considerable interest has been aroused in recent years concerning the question of just how much foetal position determines the presence of deformities or

isting at birth. We do see on occasion a child born with what appears to be bilateral contractures of the sternocleidal mastoid muscles.

Etiology

Rarely this is due to a congenital loss or a stretching of the antagonistic posterior cervical muscles and ligaments More often, however, it appears to be a positional contracture and is associated with a generalized rounded kyphosis of the entire spine similar to foetal position.

Clinical Picture

There is a flexion deformity of the neck. Frequently the deformity is accompanied by an acute hip flexion deformity or such severe flexion that the legs lie up along the abdominal wall The head is flexed on the neck and whole of the spine presents a generalized rounded, dorsal and lumbar kyphosis.

Treatment

In such a case, gradual correction by traction, starting in a line of deformity with the child on a hard bed and using pads or pillows to conform to the deformities, and then gradually decreasing these until the spine and head is flat, brings about complete correction. If the legs are involved the line of traction is gradually shifted Correction is not sufficient and treatment should not cease until a normal lumbar curve has been obtained and the head can be easily hyperextended Many of these minor contractures can be overcome merely by manual manipulation Maintenance of the corrected position for a period may be necessary to allow overstretched muscles to become relatively shortened with growth of the skeleton.

MUSCULAR TORTICOLLIS

Muscular torticollis is often termed "congenital torticollis This form of wry neck has a characteristic deformity Its etiology and pathology as well as its treatment have been a subject of considerable controversy and confusion over the years. The sternocleidomastoid muscle is the primary structure involved.

Etiology

Many theories as to the cause have been promoted but perhaps the most recently accepted is that of Chandler The intrauterine theory regards the condition as caused by abnormal pressure position, or trauma to the head neck and particularly the sternocleidomastoid muscles during intrauterine life. The hereditary theory suggests a hereditary abnormal defect in the formation of the sternocleidomastoid muscle. The infectious theory based on pathologic specimens resembling infectious myositis suggested an infection either intrauterine or in a muscle damaged at delivery The "birth trauma' theory had probably the widest acceptance It was believed that the sternocleidomastoid muscle was ruptured by some birth trauma and a hematoma developed. This hematoma" was invaded by fibrin and then followed by fibroblasts to form scar tissue. It was felt that this caused the shortening of the muscle and subsequent deformity

"Arterial occlusion" and subsequent ischemia of muscle seemed an excellent explanation, but does not fit the pathologic picture

"Venous occlusion" during delivery has been a prevailing theory but Chandler believes it to be an incomplete explanation

On the basis of anatomic and pathologic dissections, Chandler has developed a theory which seems most generally acceptable. This theory embraces the idea of intrauterine malposition with its resulting pressure on and ischemia of the sternocleidomastoid muscle which makes it atrophic, maldeveloped, ischemic, and shortened Such forces as intrauterine malposition might also be a cause of breech presentation which is present in about fifty per cent of the cases. In the other fifty per cent there is usually a history of abnormal delivery The atrophy and ischemia of the muscle make it liable to damage and breech delivery, or any abnormal delivery adds trauma at birth to this already damaged muscle

Pathology

The tumors in muscular torticollis wherein the sternocleidomastoid muscle is involved are fusiform masses involving all or part of the muscle belly usually in the middle third. The tumor is limited by "perimysium" and may extend proximally or distally to include the entire muscle It is firm and separates easily from surrounding tissues In specimens studied by Chandler and Altenberry there were no signs of blood clot formation or residual blood pigment Microscopically in all stages even in the earliest stages degeneration of muscle fibers is seen Fibrous tissue replaces many normal muscle fibers, forming a hyperplastic fibrous mass.

Clinical Picture

The mass in the sternocleidomastoid muscle may be evident at birth, but most frequently it is not noticed until after approximately ten days to two weeks, and then as a hard, immobile, fusiform swelling in the muscle Delivery has usually been difficult or a "breech presentation" The swelling increases in size for two to four weeks and then gradually subsides over a five- to eight month period.

The deformity is produced by contracture of the involved muscle the occiput being held constantly toward the involved side, the chin being rotated toward the opposite shoulder

With persistence of the deformity the skull becomes foreshortened, the level of the eyes change the side of the face on the involved side becomes flattened and the mastoid process becomes more prominent The clavicle and shoulder become elevated as compared to the opposite normal side With continued growth and no treatment a lower cervical upper dorsal scoliosis will develop

Treatment

When recognized early after birth, deformity can be corrected by gentle, yet even and persistent stretching to an overcorrected position *i e*, the head is stretched over toward the opposite shoulder from the side of involvement and the chin is rotated toward the side of involvement. Many useful tricks and



Figure 190 Posterior view—muscular torticollis in older child showing tilt and elevation of shoulder on affected side.

artifices may be used to stimulate the child to turn the head toward the involved side The crib may be placed so that the child must turn to the desired position of overcorrection in looking for window light or a favorite rattle. Special skull caps have been used with an arm attachment so that weight and use of the arm will pull the head over Placing of sandbags along the head has been found useful.

Removal of the tumor is not necessary and has the disadvantage of deforming the cosmetic outline of the neck. The rare case which has not achieved full correction on exercises can have the muscle lengthened in its sternal portion and divided at the origin from the mastoid process Such a procedure preserves the neck outline

In older patients correction of deformity can usually be obtained by lengthening of the sternal and detachment of the clavicular head of the sternocleidal muscle with an upper pole myotomy in addition, if necessary The overcorrected position can be maintained by a special collar

ROTARY SUBLUXATIONS

An inability to rotate the head may occur as an acute episode in children undergoing some injury though minor in the course of play There is usually some history of trauma mild or severe and the child suddenly develops an inability to rotate the neck to one side A sore throat or upper respiratory infec-



Figure 191 Muscular torticollis becoming more evident with growth and involving primarily the clavicular portion of the sterno-cleidal mastoid.

tion may also precede the symptoms in some cases. The resulting "wry neck" or torticollis is characterized by spasm and tenderness on the long side of the torticollis instead of on the short side as we see in such conditions as myositis. There is also limitation of motion toward the long or stretched side. Stimson and Swenson have stated that this acquired deformity is the result of a unilateral subluxation without associated fracture of the second cervical vertebra on the third. A. B Ferguson Sr has described a rotary subluxation causing such a deformity between the atlas and the axis In a series of cases of our own we were able to demonstrate that subluxation occurred in both places simultaneously in this deformity there being a subluxation between C-1 and C-2 as well as between C-2 and C-3 It is the opinion of many that the reason these subluxations occur in childhood is because of the slope of the articular facets which gradually change angle until the more stable adult situation is reached The subluxation occurs in the upper three vertebrae There is a more horizontal slope to the facets allowing sufficient rotation



Figure 108 Fibroplastic process in congenital torticollis. The dark area in the upper right portion of the section represents calcium deposition ×140

Clinical Picture

Following a respiratory infection or some injury at play or in connection with muscle spasm as the result of a cervical gland infection a child will complain of pain and inability to move the head The head is tilted to one side and can be pushed further toward that side without pain The rotation is limited to one side and when acute lateral tilt is also limited Muscle spasms and tenderness are also evident on the long or stretched side. There may also be tenderness of the spines of the first and second cervical vertebrae.

Pathology

The pathology can be demonstrated by roentgenogram. An open mouth antero-posterior view of the first and second cervical vertebrae is necessary for



Figure 193 Open mouth view of cervical spine in patient with muscular torticollis revealing tilt but accurate alignment of chin spine of C2 and base of odontoid with no evidence of cervical subluxation.



Figure 194 Cervical subluxation at C1 and C2 evidenced by malalignment of axis of C2 (base of odontoid and spine of C2) in relation to chin (correlating with C1)

diagnosis. The symphysis of the mandible, the body of the second cervical and the spine of the second cervical lie in a constant relative position, one to the other If a true antero-posterior view is taken, they will all lie on the same line If the view is slightly oblique, they will all be displaced in relative proportion to each other If, however a rotary subluxation has taken place, one of these points will be out of line in relation to the other two The mandible is carried to one side since its position will be associated with the first cervical vertebra. *The body of the second cervical and the spine give a line indicating the plane* of rotation which will deviate from that indicated by the mandible. In the lateral stereo view the unilateral subluxation between the second and third cervical vertebra can be seen the facet of the second cervical riding upward and forward on its mate below without complete dislocation.

Treatment

Reduction may occur quite easily and quickly through the use of cervical traction, and often within twenty four to forty-eight hours the neck is freely moveable A modified Thomas collar may be used thereafter for a period to prevent recurrence This would usually mean until all tenderness and any respiratory infection has subsided.

FASCITIS AND MYOSITIS

Etiology

Inflammatory conditions of the muscles or fascia usually result from exposure of the neck to drafts or dampness. It may represent a response to some focus of infection elsewhere in the body particularly if there has been an injury to the muscle.

Clinical Picture

The patient has pain on palpation of the involved muscle and spasm of this muscle will pull the head into a wry neck position. Pain and tenderness therefore are on the concave side of the torticollis and motion is limited to the convex or stretched or unaffected side. The reverse is true in unilateral subluxations.

Treatment

Heat is effective in helping quiet the inflamed muscle, fascia or tendon. Gradual traction may be necessary starting out in the line of deformity in *order to overcome the spasm.* Immobilization with a collar is then advisable until all tenderness has subsided

ARTHRITIS

Atrophic Ankylosing (Still s Disease) It is perfectly true that older children can be afflicted with a mixed or rheumatoid type of arthritis, but in childhood—as a rule—one meets most often with that generalized debilitating, malignant, ankylosing desperate affliction known as Still s Disease.

Etiology

Dimson believes the condition to actually be a subdivision of rheumatoid arthritis It is seen mostly in girls, beginning around three to five years of age, and often seems to follow some infectious illness In the early stages one must be careful to differentiate between acute and rheumatic fever, or later those joint manifestations which occur in the recovery stage of rheumatic fever, sometimes termed chronic, fibrous rheumatism of Jacoud

Clinical Picture

Painful swollen joints accompanied by fever, tachycardia starting rather abruptly and accompanied by lymphadenitis, splenomegaly, leukocytosis, anemia and cachexia constitute the picture of this disease. The sedimentation rate is elevated

In these children there seems to be a predilection for the cervical spine as well as small and medium sized joints. The neck becomes immobile and flexed A peculiar, birdlike facies and a waxy pallor develops insidiously

Treatment

Treatment of the patient as a whole is of course essential in order to remove the foci of infection and correct the anemia and cachexia. Deformities in all joints should be prevented as correction of deformities once acquired is difficult The cervical spine should be supported by a Thomas frame and headpiece or the normal cervical curve retained or reobtained by cervical traction and a neck roll Active exercise within the limits of pain is important in maintaining motion.

In severe quiescent cases operative correction of deformities may be necessary

Cortisone and its derivatives, other than often dramatically relieving pain have been disappointing. The drug may even give relief of all joint signs and symptoms but only as long as treatment is continued. When attempting to taper off the dosage relapses tend to occur Prolongation of treatment causes many undesirable side effects. Indeed this condition is one of the unhappiest of childhood afflictions.

Intra articular hydrocortone therapy has seemed to help a few

References

HARTLEY J B AND BURNETT C W F Spina bifida and its associated skull defects Arch Dis Childhood 18: 173 1943

PICK, DAVID M Spina bifida and its associated malformations. Wisconsin M J 50: No 7 July 1951 659–663.

TAYLOR, J C Breech presentation of hyperextension of the neck and intrauterine dislocation of cervical vertebrae. Am J Obst. & Gynec. 56: 381 1948

HORWITZ, A E. Congenital elevation of scapula. Am. J Orthop Surg 6: 260 1908.

SCHROCK, R D Congenital elevation of scapula. J Bone & Joint Surg., 8: 207 1926

SMITH A DeF Congenital elevation of scapula. Arch. Surg 42: 529 1941

JEANOPOLOUS C J Congenital elevation of the scapula J Bone & Joint Surg 34A: 883, 1952

GESELL, A Hemihypertrophy and mental defect. Arch Neurol & Psychiat 6: 400 1921

GESELL, A. Hemihypertrophy and mental defect. Arch. Neurol & Psychiat., 6: 400 1921.
WARD J AND LEARNER, H. H. A review of the subject of congenital hemihypertrophy and a complete case report. J Pediat., 31: 403 1947
NELSON W E. Skeletal defects. In Textbook of Pediatrics, 5th ed., 1449 W B Saunders, Philadelphia, 1950
CAMPBELL'S "Operative Orthopedics" Vol. 11 Mosby St. Louis, 1949
COMPERE, E. L. Excision of hemivertebrae for correction of congenital scoliosis. J Bone & Joint Surg 14: 555 1932
LEWIS W H The development of the arm in man. Am J Anat., I: 145 183 1901–1902.
KEITH A Human embryology and morphology 6th ed., The Williams & Wilkins Co Baltimore 1933
SHOUL, M I AND RITRO, M Manifestations of Klippel-Feil syndrome Am. J Roentgenol. 68: 369 1952.
SUTRA, C. J AND MISHKIN R D Familial nuchal rigidity form of hypomobility of vertebral column which clinically may be misdiagnosed as ankylosing arthritis (antithesis to Ehler Danlos disease) Bull. Hosp Joint Dis., 13: 155 1952.
CHANDLER F A. AND ALTENBERG, A Congenital muscluar torticollis. J.A.M.A. 125: 476, 1944
CHANDLER, F A Muscular torticollis. J Bone & Joint Surg 30A: 566 1948.
STIMSON A B. AND SWENSON PAUL C. Unilateral subluxation of the cervical vertebrae without associated fracture J A. M A 104: 1578, 1935
EASTWOOD W J Discussion on fractures and dislocations of cervical vertebrae. Proceedings of the royal society of medicine 33: 1940
ROBERTS S M Fracture and dislocations of the cervical spine. J Bone & Joint Surg., 19: 1937
BERKHEISER, E. J AND SEIDLER, FERDINAND Non-traumatic dislocations of the atlanto-axial joint. J A. M A., 95: 517 1931
COUTTS M D Atlanto-epiphyseal subluxation. Arch. Surg., 29: 297 1934.
JONES R. W Spontaneous hyperemic dislocations of the atlas. Proc. Roy Soc. Med., 25: 1 1931–32
PATTON E. F Cervical subluxation. Ann. West. Med. & Surg., 3: 1949
BIRKETT A N Fractures and dislocations of the cervical spine In Modern Trends in Orthopedics, ed. by Harry Platt, 293 Hoeber New York, 1950
TRAVELLI L. Subluxations of the cervical spine following acute rhino-pharyngitis. Minerva Med., 1947
NICHOLSON J T Spontaneous reduction of cervical spine dislocations. J A M A., 115: 2063 1940
TOWNSEND E. H JR. AND ROWE, M L Mobility of the upper cervical spine in health and disease. J Pediat., Vol. 10 #5 November 1952, 567–574.
JACOBSON G AND ADLER, DENNIS O Evaluation of lateral atlanto-axial displacement in injuries of the cervical spine Radiology 61: 1953
PETERS K. et al., Handbuch der Anatomie des Kindes, 2: 1938.
HERZ, E AND HOEFER, P F A. Physiologic analysis of involuntary motor activity Arch Neurol & Psychiat., 61: 129 1949
WYCIS, H T AND MOORE, J R. The surgical treatment of spasmodic torticollis. J Bone & Joint Surg 36A: 119 1954
HOWORTH M B Spasmodic torticollis. Textbook of Orthopedics, W B Saunders, Philadelphia, 1063 1952
KAPLAN E. B Personal communication.
SHANDS, A. R. Affections of neck and shoulder Handbook of Orthopedic Surgery Ed. 2, 434, C V Mosby Co St. Louis, 1940
COMPERE, E. L. Neck, shoulder and arm syndrome. New Orleans M & S. J 104: 473, 1952.
COAKLEY W A TEPLITSKY D AND DIAZ, A. Infections of head and neck. Am. J Surg. 77: 460 1949
MERCER, R. D Atlas-axis dislocation following cervical adenitis. Cleveland Clin. Quart., 11: 161 1952
DIMSON S. B. The diagnosis and treatment of Still's disease. Rheumatism 10: 18 1951.

6

The Upper Extremity

FRANK STELLING M.D

The hand of the child is fundamentally no different from the hand of the adult in its response to injury, healing characteristics and therapeutic principles. There are certain basic differences which should be pointed out. In the adult, the major cause of disability is trauma because of the preponderance of industrial accidents In the child the major disabilities are caused by paralytic conditions and congenital abnormalities, with trauma being less frequent but certainly no less important The same sound surgical principles apply to the child as well as to the adult, however, techniques must be varied to fit the individual problem. There are those conditions such as the spastic hand and congenital abnormalities that should be approached differently in the child than the adult. Some problems encountered in childhood must be treated conservatively with therapy directed toward prevention of deformity until maturity has been reached, before definitive procedures are performed. Occasionally, time consuming reconstructive procedures may be carried out in children that would not be attempted in adults because of the time factor and the economic impossibility

This chapter will deal mainly with the discussion of those conditions common to children and the application of sound therapeutic principles. No attempt will be made to deal with functional anatomy or the many basic fundamentals of hand surgery, all of which are most important and absolutely necessary in order to carry out the procedures to be described.

CONGENITAL ANOMALIES

Congenital anomalies of all types account for about 25 to 30 per cent of all the admissions to children's hospital services throughout the United States Deformities of the hand comprise 20 to 30 per cent of this group No single deformity can be considered common and some are extremely rare and need not be mentioned in this section. Most of the deformities are mixed in nature, and in nearly every instance treatment must be individualized. In spite of this, certain general principles of treatment for the more common groups of anomalies will be considered here.

The etiology of congenital anomalies of the hand has not been definitely

established. A discussion of the theories which have been advanced will not be considered here, but it would seem that the most acceptable theory is that of mutations which are subsequently inherited.

CLUBHAND OR CLINOARTHROSIS

Clubhand or clinoarthrosis are terms denoting a deformity of the hand manifested by radial deviation resulting from a defect in the radius. This defect may be either a complete or partial absence of the bone This condition is a manifestation of a defect in the radial elements of the limb bud. Similarly an ulnar clubhand may occur in association with an ulnar defect. It has been found that the dividing line between radial and ulnar hemimelias lies along the index finger Quite often the thumb is absent in those radial clubhands with a defect in the radius while often the last three fingers are absent in the defects in the ulna.

Associated Anomalies

The skeletal changes which occur in the upper extremity usually involve most of the bones The scapula is commonly reduced in size and the clavicle may be shorter The humerus is usually shorter than normal and either end may present some deformity The carpal bones are rarely complete in number The one most frequently absent is the navicular, followed by the greater multangular Quite frequently the first metacarpal is absent. Varying combinations occur in regard to the relationship of the first metacarpal and the thumb If the first metacarpal is missing the thumb usually has no function and very often is a soft tissue appendage

Muscular Defects

Numerous defects in the muscles accompany the condition. If the thumb and first metacarpal are present, the extensor pollicis longus and brevis and abductor brevis are also present and the muscles of the thenar eminence are usually normal. If the first metacarpal is absent, however these muscles are commonly missing. The pronator teres may be missing if the radius is completely absent or it may be underdeveloped if there is a partial defect in the radius. The flexor pollicis longus is apt to be absent unless the entire thumb and first metacarpal are present in which case it is likely to be normal. The muscles about the shoulder and arm are frequently involved. The pectoralis major may have an abnormal insertion or its clavicular or costal portions may be absent. The pectoralis minor and deltoid are usually present although the latter muscle may have an abnormal insertion or it may fuse with the triceps or the brachialis. The biceps is frequently missing however if it is present in a patient with a complete absence of the radius, it inserts into the lacertus fibrosus. The brachialis varies but is usually present and the triceps is usually normal. The brachioradialis is found to be missing when the radius is completely absent however occasionally it will be present and inserted into the ulna. The extensor carpi radialis longus and brevis may be absent or they may be fused with the extensor digitorum communis which is usually present and normal The supinator is commonly absent unless the proximal radius is present.

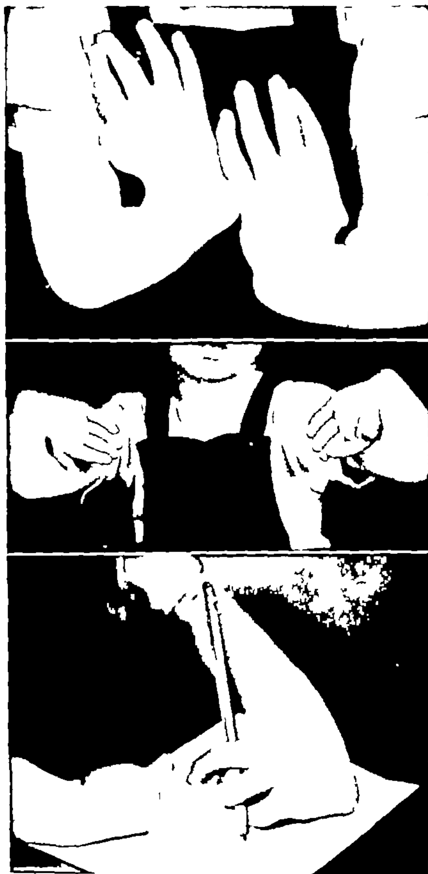


Figure 195 Typical clubhand bilateral with poor development of the thumbs

established. A discussion of the theories which have been advanced will not be considered here, but it would seem that the most acceptable theory is that of mutations which are subsequently inherited.

CLUBHAND OR CLINOARTHROSIS

Clubhand or clinoarthrosis are terms denoting a deformity of the hand manifested by radial deviation resulting from a defect in the radius. This defect may be either a complete or partial absence of the bone. This condition is a manifestation of a defect in the radial elements of the limb bud. Similarly, an ulnar clubhand may occur in association with an ulnar defect. It has been found that the dividing line between radial and ulnar hemimelias lies along the index finger. Quite often the thumb is absent in those radial clubhands with a defect in the radius, while often the last three fingers are absent in the defects in the ulna.

Associated Anomalies

The skeletal changes which occur in the upper extremity usually involve most of the bones. The scapula is commonly reduced in size and the clavicle may be shorter. The humerus is usually shorter than normal, and either end may present some deformity. The carpal bones are rarely complete in number. The one most frequently absent is the navicular, followed by the greater multangular. Quite frequently the first metacarpal is absent. Varying combinations occur in regard to the relationship of the first metacarpal and the thumb. If the first metacarpal is missing, the thumb usually has no function and very often is a soft tissue appendage.

Muscular Defects

Numerous defects in the muscles accompany the condition. If the thumb and first metacarpal are present, the extensor pollicis longus and brevis and abductor brevis are also present, and the muscles of the thenar eminence are usually normal. If the first metacarpal is absent, however, these muscles are commonly missing. The pronator teres may be missing if the radius is completely absent or it may be underdeveloped if there is a partial defect in the radius. The flexor pollicis longus is apt to be absent unless the entire thumb and first metacarpal are present, in which case it is likely to be normal. The muscles about the shoulder and arm are frequently involved. The pectoralis major may have an abnormal insertion or its clavicular or costal portions may be absent. The pectoralis minor and deltoid are usually present, although the latter muscle may have an abnormal insertion or it may fuse with the triceps or the brachialis. The biceps is frequently missing; however, if it is present in a patient with a complete absence of the radius, it inserts into the lacertus fibrosus. The brachialis varies, but is usually present, and the triceps is usually normal. The brachioradialis is found to be missing when the radius is completely absent, however, occasionally it will be present and inserted into the ulna. The extensor carpi radialis longus and brevis may be absent or they may be fused with the extensor digitorum communis, which is usually present and normal. The supinator is commonly absent unless the proximal radius is present.

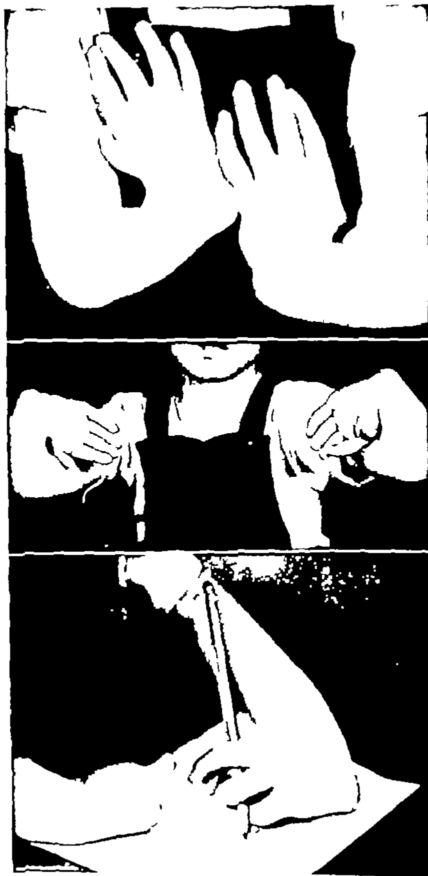


Figure 195 Typical clubhand bilateral with poor development of the thumbs



Figure 196 Clubhand with absence of the thumb

Vascular Defects

The arterial system shows numerous abnormalities. The brachial artery may be normal or it may divide into two branches high in the arm. The ulnar artery is usually normal but the radial artery is frequently small or absent. The median nerve is not infrequently the most superficial structure found on the radial aspect of the forearm. For the most part the ulnar nerve is normal and supplies the usual intrinsic muscles in the hand.

Treatment

Many different methods have been devised through the years to improve these hands. Most of the operative procedures devised for the condition have resulted in varying results from fair improvement to doubtless value. The general methods of treatment of this condition at the present time are (1) conservative stretching and splinting, and the development of the use of the hand without a surgical procedure, (2) stretching and splinting followed by tenotomy and soft tissue surgery to get the hand over the ulna, followed by fusion of the wrist or of the ulna into the carpus and (3) that of getting the hand over the wrist by conservative or surgical procedures or both followed by some bony procedure in order to keep the hand in that position without a definite stabilisation. In general the operative procedures that have been described in the literature have been confined to tenotomy of the contracted muscles, osteotomy of the curved ulna and the addition of a stabilizing bone graft to replace the absent radius or splitting of the ulna into radial and ulnar portions. Most of these procedures result in a fusion of the wrist joint, and if such is the result, the cosmetic appearance of the hand is certainly more pleasing but sometimes it is questionable if the functional value of the hand has been increased.

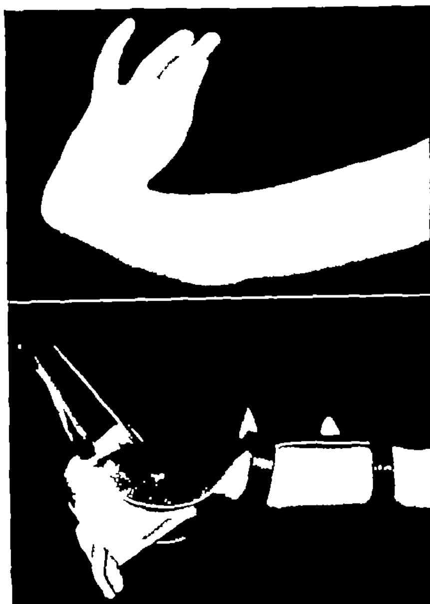


Figure 107 Typical clubhand showing the use of clubhand splint. This is used part time to give active stretch in order to lessen the deformity

Conservative Treatment

Conservative treatment should be started in all cases as early as possible. In the correction of the deformity, plaster casts splints or braces should be used until the soft tissues have been stretched to the greatest degree that the bony deformity will allow As soon as possible the deformity is corrected by plaster casts. Following this small correction braces or splints may be used part time while stretching is carried out as a daily routine by the parents. Some authorities feel that this is the only treatment that should be done. Quite often if treatment is carried on during the entire growth period definite deformity will continue but frequently the child adapts to this deformity Stretching and the movements of the parts keep the hand functionally sound and it is amazing

to see how well these children can use the hand at the termination of the growth period. Cosmetically the hand is not too pleasing but quite often, the function is very satisfactory, and perhaps better from a functional standpoint than one with a wrist fusion alone.

Surgical Treatment

Recently Riordan devised a modification of the surgical method of Starr which seems to be more promising than any other surgical procedure that has yet been devised. Starr transplanted the proximal end of the fibula to replace the distal end of the absent radius Riordan has used this method in those instances of a partially absent radius and also used the same procedure by grafting in the proximal fibula into a split portion of the shaft of the radius giving a definite support to the radial side of the carpus He stresses that first attempts should be those of conservative stretching of all the soft tissues to the maximum during the early period of life. This is followed by surgical release of the carpus, placing the bones of the hand over the distal end of the ulna. This is an important step and must be done prior to the insertion of the bone graft. He also emphasizes that this cannot be done by conservative, nonsurgical methods. He has found it nesessary to cut the fascia but in no instance has he had to tenotomize any of the musculotendinous structures. The hand is maintained in the corrective position by means of Kirschner wires and incorporated in plaster immobilization. He advises osteotomy of the ulna to correct bowing some two or three months after re-positioning of the hand if this is necessary The final step is that of the addition of a stabilizing factor to the radial side of the forearm. The proximal end of the fibula including the epiphysis is either grafted into the distal end of the partially present radius or directed obliquely against the ulna into a V-shaped notch after turning down a long spike in those instances of a complete absence of the radius. Solid fixation of the graft to the bone of the ulna is necessary, otherwise it may be absorbed. In those instances where the thumb is absent, the majority of the patients use a pinching mechanism between the index and long fingers, and one is usually tempted to swing the index finger around in order to make a thumb This can be performed but should not be done in a unilateral case. It is felt that a hand with only three normally functioning fingers and an abnormal thumb is not as good from a cosmetic standpoint or functional standpoint as four normal fingers and no thumb In a bilateral case, the thumb may be reconstructed on one side. Corrective procedures are probably best done as early as possible before abnormal motor patterns are established.

References

ALBEE, F H. Formation of radius congenitally absent condition seven years after implantation of bone graft. Ann. Surg., 87: 105 1928

BUNNELL, S Surgery of the Hand. J B Lippincott Co Philadelphia, 1944

KATO, KATSUJI Congenital absence of the radius. J Bone & Joint Surg 6: 589 1924

RIORDAN D C Congenital absence of the radius. J Bone & Joint Surg 37A: 1129 1955

STARR, D E Congenital absence of the radius, a method of surgical correction. J Bone & Joint Surg., 27: 261, 1945.



Figure 108 Thumb clutched hand with finger deformities

CONGENITAL FLEXION AND ADDUCTION DEFORMITY OF THE THUMB
(POLLEX VARUS) (THUMB CLUTCHED HAND)
(CONGENITAL CLASPED HAND)

This anomaly is a rare deformity which tends to be an inherited trait. The thumb is characteristically held adducted into the hand with marked flexion of the metacarpophalangeal joint. Quite frequently there are coexistent flexion deformities of the proximal interphalangeal joints of the fingers thereby producing the term, thumb clutched hand. The skin of the flexor surface of the fingers and thumb is usually contracted. This is thought to be secondary to the true pathology The flexion deformity of the proximal interphalangeal joints of the fingers may exist without the thumb deformity, but this is considered to be a rare occurrence In most instances, it has been found that there is a hypoplasia or absence of the extensor pollicis brevis with secondary adduction flexion contracture of the thumb In the finger deformities, it is believed that there is a hypoplasia or absence of the intrinsic musculature.

Treatment

The results of treatment depend upon the stage in which therapy is started Many of these cases have been noted in adults with persistent disabling finger and thumb deformities. If the deformity is seen early, conservative measures should be instituted. The thumb should be mobilized out of the palm into extension and abduction, holding them in this position by some type of splint. White and Jensen use a special elastic abduction splint which can be easily removed for cleaning. Weckesser uses a plaster cast for several months changing the cast at four to six week intervals in order to allow for growth. Most of his cases developed good function with this conservative care. If the deformity returns after releasing the part following conservative immobilization, then it is to be expected that the tendon is either absent or nonfunctional, and tendon



Figure 100 Congenital absence of the intrinsics without clutched thumb

transplants should be done White transplants the *common extensor tendon of* the index finger into the extensor surface of the metaphysis of the base of the proximal phalanx of the thumb and has shown excellent results in these cases. Sometimes the skin is tight on the flexor surface and a Z-plasty or releasing incision with a full thickness graft should be utilized. The contracture has been limited to the skin and does not usually involve fascia and deeper structures in the small child In the adult contracture of all elements of the thumb web is to be expected.

References

WECKESSER, E C Congenital flexion-adduction deformity of the thumb (congenital "clasped thumb") *J Bone & Joint Surg.*, 37A: 977 1955

WHITE, J W AND JENSEN W E. The infant's persistent thumb clutched hand. J Bone & Joint Surg 34A: 680 1952.

ZADEK, I. Congenital absence of extensor pollicis longus of both thumbs, operation and cure. J Bone & Joint Surg 16: 432, 1934

ZUIDEMA BARRETT Congenital symmetrical finger contractures. J A M A., 155(5): 437 1954.

STENOSING TENOVAGINITIS OF THE THUMB—"TRIGGER THUMB

This condition is characterized by a flexion deformity of the interphalangeal joint of the thumb and occurs in adults as well as in children. In the adult, it is irritative in origin due to repeated trauma and in the child it is congenital. Snapping is usual in adults and rare in children. *It is generally felt that* the lesion is of congenital origin in infants even though in some instances it is not present until weeks or months after birth. Zadek cites a case of an infant with a history of minor trauma of one thumb that was thought to have developed as a result of the injury however a few months later, the other thumb developed the same deformity with no history of trauma The pathology was found to be the same in both cases.

Pathology

The gross pathological appearance of this lesion is that of an encircling constricting fibrocartilaginous band of the sheath of the flexor pollicis longus at the of the metacarpophalangeal joint. Usually as this constriction is released there is an indentation or groove in the tendon with thickening on either side of this indentation Sometimes the tendon is bulging to double its normal size on either side of the groove

Clinical Picture

The interphalangeal joint of the thumb is held in a position of fixed flexion. There is inability to actively extend the joint. Usually the thumb can be flexed actively a few degrees In some instances the distal joint can be forcibly extended and then locks in flexion as soon as flexion is again initiated. The joint cannot be passively extended and any attempt to do so causes pain As extension of the joint is attempted the tendon becomes taut distal to the metacarpophalangeal joint, and the skin of the flexor surface of the joint blanches There is a thickened area on the flexor surface of the thumb at the level of the metacarpophalangeal joint This area may at times be sensitive.

Treatment

If the interphalangeal joint can be passively extended and splinted in this position for three weeks, the thumb sometimes resumes normal function with permanent loss of the deformity In most instances, however, this treatment is unsuccessful and the deformity recurs The treatment of choice is that of incision of the flexor tendon sheath, preferably by making two incisions through

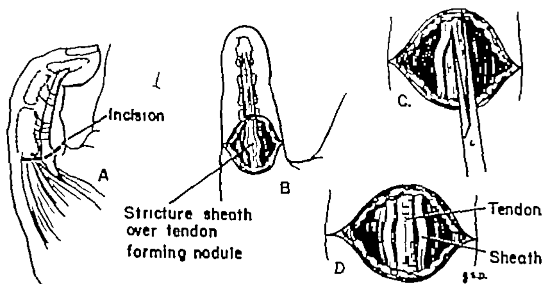


Figure 200 Operation for "trigger thumb" The incision in the skin is transverse and the incision in the tendon sheath longitudinal.

each side of the annular constricting band and by removing a small section of the sheath. The incision in the skin should be made transversely in a line with the crease at the metacarpophalangeal joint. A longitudinal incision crossing the skin crease will produce a serious painful scar which can produce a flexion deformity Care must be taken not to injure the digital nerves. The results of this therapy are usually excellent.

References

FAHEY J J AND BOLLINGER J A. "Trigger thumb" in adults and children. J Bone & Joint Surg., 36A: 1200 1954.

JAHSS S A Trigger finger in children. J.A.M.A 107: 1463 1936

MILLER, J W Pollex varus, a report of two cases Univ Hosp Bulletin Ann Arbor Michigan, 10: 1 1944.

SPRECHER, E. E. "Trigger thumb" in infants. J Bone & Joint Surg., 31A: 672, 1949

WHITE, J W AND JENSEN W E. "Trigger thumb in infants. Am J Dis. Child., 85: 141, 1953.

ZADEK, ISADORE Stenosing tendovaginitis of the thumb in infants. J Bone & Joint Surg 24: 326 1942

SYNDACTYLISM

Syndactylism is the most frequent anomaly of the hand and occurs with all degrees of severity The simplest involvement is an incomplete web formation between the two fingers with no bone or joint deformity Extreme involvement may exist in the form of severe webbing of all the fingers with incomplete segmentation of the bony parts and multiple joint deformities There will occasionally be coexistent symphalangism or extra bony segments may be interspersed. Syndactylism is often associated with polydactylia or brachydactylia or a combination of all of these conditions It is frequently bilateral and is sometimes seen in conjunction with webbing of the toes or other deformities of the foot. The deformity is most frequently thought to be inherited. In the series of cases reviewed at the Shriners' Hospital in Greenville almost all of the cases have been found to originate from a single family

Treatment

The results of therapy vary directly with the degree of deformity number of fingers involved and the presence of bony fusion. Partial webbing of the skin without bone or nail involvement will result in a normal finger The presence of bone or joint deformities will invariably result in some permanent deformity in spite of the best operative procedures

Results will be most successful if the surgery is delayed until the child is four or five years of age. This rule should be abandoned, however if the growth of the finger is being held back because of a bony attachment to a shorter adjacent finger It is essential that the fingers be well immobilized postoperatively, and it is almost impossible to secure good immobilization in infants.

Many different types of procedures have been advocated but the operation of choice is that of a crossed V shaped flap at the base of the web space. The use of a single V allows for longitudinal scarring which extends up one or both of the fingers at the apex. The use of two V's crossed produces Z-plasty effect,

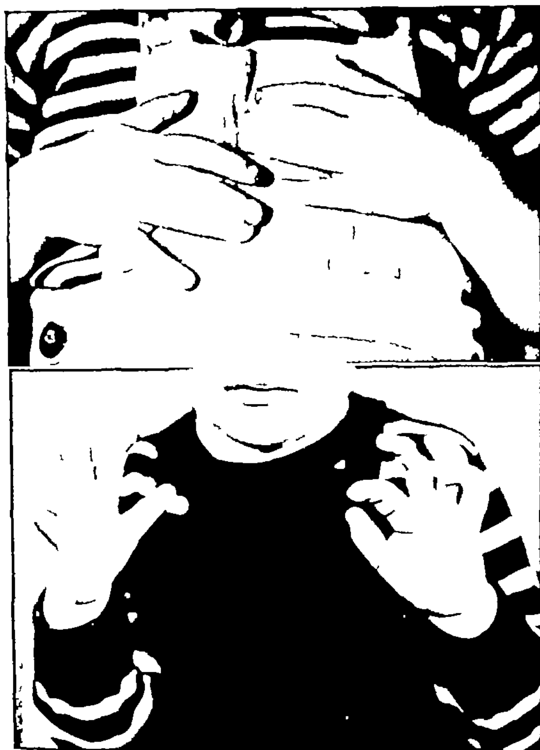


Figure 201 Syndactylism bilateral involving two fingers. No bone joint or nail deformity This child is only two years of age and a good result should be expected *Figure 202* Same case as Figure 201. Good functional and cosmetic result following surgery

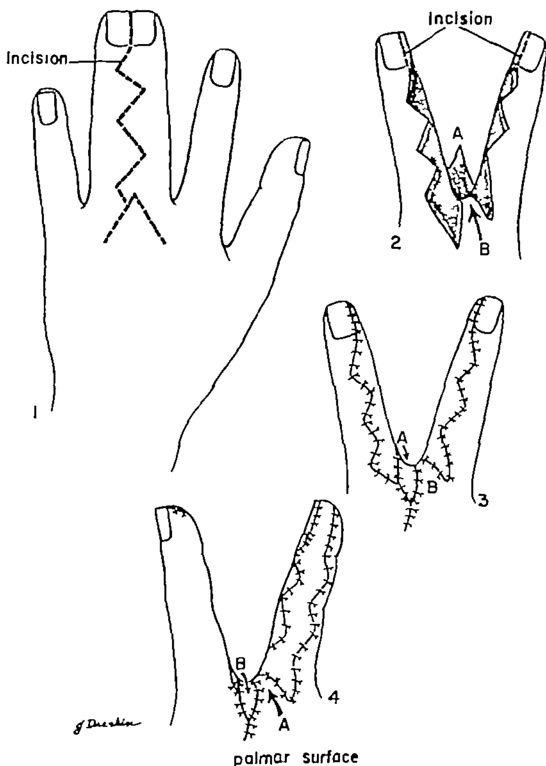


Figure 203 Method of surgical treatment for syndactylism. The sides of the finger are covered with split thickness grafts—the base by crossing two triangular flaps.





Figures 204, 205 Syndactylism involving three fingers. Note the deformity of the nails and deformity of the longer fingers. There is also lack of segmentation of the distal phalanges. Good result to be expected here but there will probably be some fixed joint contracture of the middle finger Should have been treated surgically at a younger age for a better result.

and longitudinal scarring is minimized The pedicle of these flaps should be based proximally at about the level of the metacarpophalangeal joint. The web is then separated on both sides It is essential that this web separation be irregular in order to prevent longitudinal scarring at the graft attachment. The soft tissues are separated by blunt dissection after the skin incisions are completed and the neurovascular bundles should be carefully protected. The fused phalanges should be separated with a bone cutter and trimmed smoothly If a



Figure 206 Brachydactylia, syndactylism and symphalangism. Can be improved by separating the index and adjacent finger. Will still be restricted in motion in the middle finger joints.

Figure 207 Bilateral syndactylism of three fingers plus extra digits on the ulna side. *Figure 208* Same case as Figure 207 after surgical release of the syndactylism and removal of the entire ray on the left and digit on the right.

double nail exists it should be split and a portion of the nail and its matrix on each side should be removed so that the graft may be brought around the nail margin. The resultant defect produced on the opposing sides of each finger should be covered by a free full thickness skin graft A pressure dressing is applied with the fingers in complete extension and wide abduction Further immobilization is accomplished by the use of a cast extending above the elbow and incorporating the fingers The operation is best performed under a tourniquet, but it should be released prior to the application of the full thickness grafts and the suture of the flaps The cast is removed in about ten days, and the wounds are dressed, but some form of splinting or immobilization must be continued for a six weeks' period. Joint motion is not interfered with permanently in these children in spite of the seemingly long immobilization period Both sides of any one finger should never be operated upon at one time because of the danger of losing the finger

The deformity may be such that a part is so deformed that even though the fingers are separated, function is still very poor, and there is interference, therefore, with the good function of the remaining part of the hand. In such instances it is better to sacrifice the poorly developed part and thereby gain improved function in the remaining good segments

POLYDACTYLISM

Polydactylism is inherited and often accompanies syndactylism, brachydactylism and other congenital anomalies It is probably the most common congenital anomaly, but because some of these small finger appendages attached by a fine skin pedicle are tied off with a suture in the nursery by the obstetrician or pediatrician they are lost in statistical analyses.

The anomaly occurs in three main types (1) an extra soft tissue mass not adherent to the skeleton and frequently without bone, joints or tendons, (2) duplication of a digit containing all elements and attached to a large metacarpal head or to a bifurcated metacarpal (3) a complete extra digit with a full meta carpal.

Treatment

Those cases belonging to Group 1 which show a very small pedicle at birth can be easily treated by tying off the pedicle with a piece of silk. This usually results in a normal appearing hand without deformity Sometimes the soft tissue attachment is too thick for this type of treatment, and in such cases, it is best to wait until the child is older (four to five years) before carrying out a surgical resection. Extra digits do not cause functional difficulty in the early years of life. In some instances, it is difficult to decide which digit should be sacrificed thereby making necessary careful study and observation over a period of time before making the decision to operate. In the case of thumb involvement, one should be extremely vigilant to see that tendinous attachments are preserved. Occasionally, the tendons of an accessory digit may be transferred to provide function in the remaining digit

The bifid thumb in which the duplication does not proceed beyond the termi

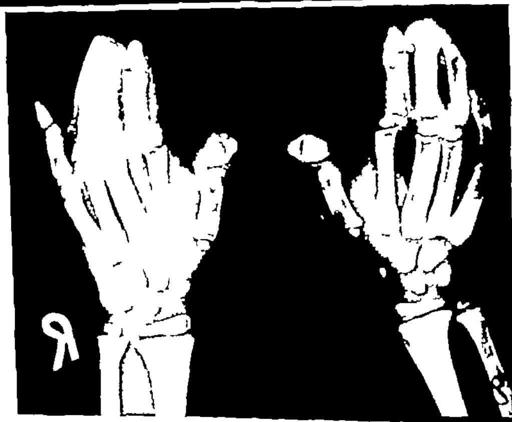


Figure 209 Bilateral syndactylism of all four fingers and bifid thumbs



Figure 210 Same case as Figure 209 after correction of the syndactylism and removal of the radial thumbs There is some residual scarring on the flexor surface but functional ability is excellent.





Figures 211 212 Severe syndactylism with only deformity of the middle and ring fingers. Wedged phalanx in the thumb Result here will not be excellent but fingers will be greatly improved.

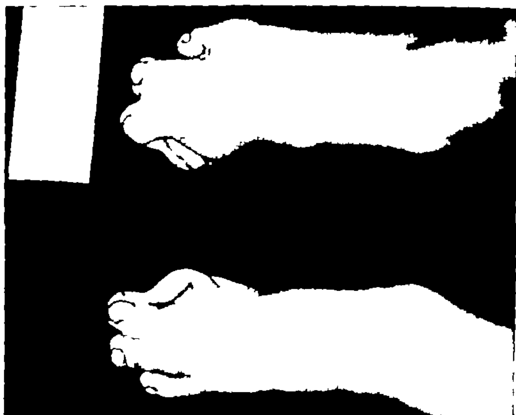


Figure 213 Same case as Figures 211 212 Deformities of the feet

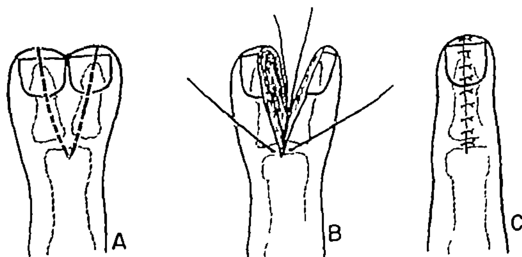


Figure 214 Surgical correction of bifid distal phalanx



Figure 215 Bilateral syndactylism of two fingers. No bone or nail deformity Child is young and excellent cosmetic and functional result should be anticipated with proper surgery



Figure 216 Same case as Figure 215 Postoperative appearance Good functional result

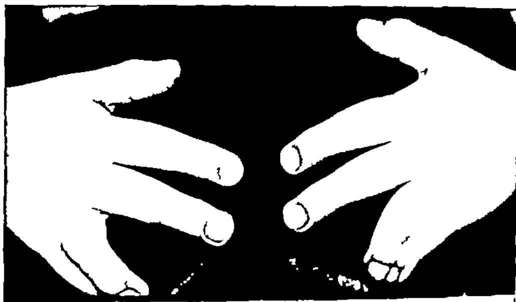


Figure 217 Syndactylism of two fingers bilateral. Some nail deformity and some joint deformity



Figure 218 Same case as Figure 217 Surgery performed early enough for joint deformity to be corrected.



Figure 219 Congenital anomalous development of joint in fingers. Large double first metacarpal of the middle finger with marked joint contracture.

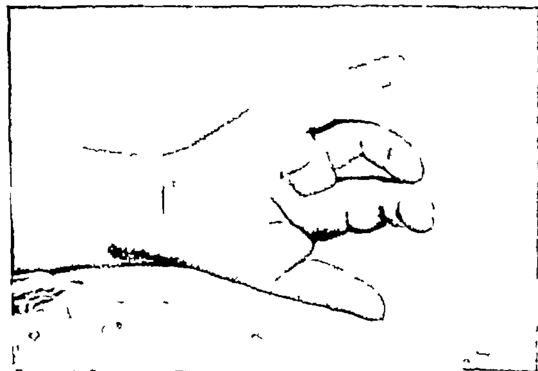


Figure 220 Same case as Figure 219 after complete ray excision and narrowing of the hand Child has very good function.

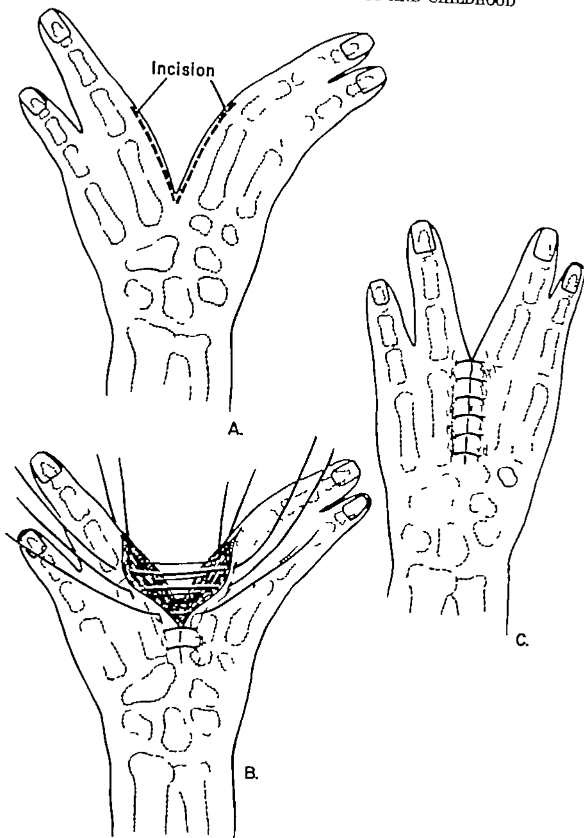


Figure 2.1 Narrowing of hand to improve cosmetic appearance. Function should always be considered however



Figure 222 Extra ulnar digits



Figure 223 Same case as Figure 222 Final result after surgical excision



Figure 224 Bifid thumb Excision of the radial digit will give excellent result

nal phalanx is a form of polydactylism. In these cases an excision of the central wedge from each tip and joining the resulting fragments is the procedure of choice

CLEFT HAND

This is a rare deformity with many variations and many other types of congenital malformations may occur concomitantly such as syndactylism brachydactylism and symphalangism. The usual deformity is that of a central cleft dividing the hand in two parts. The digits in each part may be webbed partially or completely or they may be free. Most frequently, one or more rays are missing. In some cases there may be a radial and ulnar ray with no digits between. In others the hand may be normal except for the lack of a central digit. In nearly all instances the patient uses the part almost like a claw opposing one part to the other for grasping.

Treatment

The treatment must be individualized to the type of deformity present. In those instances where there are only two functioning units present and no webbing, it is best that the hand be left untreated

The presence of webbing should be released as previously described under syndactylism and the cleft eliminated by closure of the palm and hand defect.



Figure 225 Lobster claw hand No treatment necessary in this type case

If a central digit or digits are missing but their metacarpals are present, the metacarpal should be excised and the two parts of the hand closed

MEGALODACTYLISM

Hypertrophy of one or more digits or the entire hand may occur as a rare anomaly These hypertrophies may be the result of true anomalies of development but there is definite evidence that a large number of these cases have neurofibromatosis as the underlying pathology MacCarroll Moore and Brooks have shown cases of increase in length as well as breadth or of soft tissues and bone due to neurofibromatous changes in the digit. The neurofibroma itself is usually unobtrusive and the associated hypertrophy is the most notable condition

Treatment

A good deal of the enlargement may consist entirely of soft tissue hypertrophy in which are located large tortuous nerves and nodular tumor masses. When this occurs the resection of the soft tissue masses and grossly enlarged tortuous nerves and redundant skin may produce a nearly normal finger although some diminished sensation may be present. It is advisable to perform this procedure in stages to avoid circulatory damage to the entire part If digital nerve is uninvolved, it should be preserved and excess tissues removed from its periphery In some instances the length of the finger and associated bony deformity is too great and parts of the finger may have to be sacrificed If the finger is severely deformed and enlarged producing a poorly functioning unit in the presence of an



Figure 226 Congenital hypertrophy Megalodactylia



Figure 227 Congenital hypertrophy or megalodactylia

otherwise normally functioning hand, it is probably best to sacrifice it. A ray amputation in these cases will produce an excellent functional and cosmetic result

ANNULAR GROOVES AND CONGENITAL AMPUTATIONS

These ring like bands appear circumferentially about the arm, forearm, leg, fingers and toes. The exact etiology is unknown, however, they are not due to amniotic bands The grooves may be shallow, involving only skin and subcutaneous tissue, or they may extend through the fascia to any depth The deeper ones often interfere with lymphatic and venous return causing distal edema At times, that portion of the extremity distal to the groove will be lost. This definitely occurs *in utero* and amputations are then present in the newborn Single or multiple amputations of fingers or the hand frequently accompany these annular grooves

Treatment

In those cases of superficial grooving no treatment is necessary and the groove should be left alone Deeper grooves and particularly those causing interference with lymphatic drainage should be corrected. The groove is excised down to normal structures releasing the constricting band, but in closing the scar should be zigzagged in approximating the subcutaneous tissue and skin in order to avoid the occurrence of a constricting scar The entire annular groove should not be excised in one procedure but it should be carried out in stages in order to avoid circulatory embarrassment

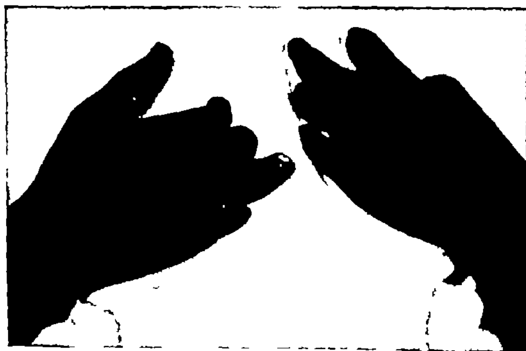


Figure 228 Congenital grooves and bands with amputations associated with partial syndactylism Some improvement cosmetically and functionally from deepening the webs.



Figure 229 Typical Streeter's dysplasia (congenital amputation and annular grooves)

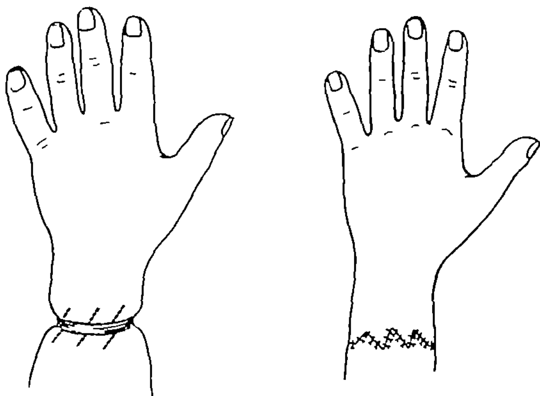


Figure 230 Correction of annular groove by surgical excision—best done in stages with one-third to one-half of circumference undertaken at one sitting

BRACHYDACTYLISM

The embryonic development may be interfered with so as to produce a shortening of one or more digits The phalanges may be decreased in number or length or the metacarpal may be shortened. Various combinations may occur simultaneously It is not uncommon to see this condition occurring concomitantly with syndactylism or polydactylism

Treatment

There is really no treatment for brachydactylia The fingers may be almost normally formed and function quite satisfactorily The accompanying deformities of syndactylism or polydactylism should be corrected as previously described

AGENESIS AND INCOMPLETE DEVELOPMENT

Absence of parts or incomplete development of parts may involve an entire extremity or parts of an extremity In some instances, the involvement may be quite extensive, even to the point of producing a quadruple amputee. The involvement may be so mild, however, as to produce a perfectly normal skeleton except with the absence of a phalanx or even a part of one phalanx. Sometimes the stump of a part is smooth with no external evidence of any part distal to it,



Figure 231 Severe partial brachydactylia and syndactylism. This child was severely upset by this deformity This would be a good case for excision of the middle and ring fingers and moving the middle ray ulnarward. The deformity would be much less noticeable.

however, by x ray there are at times evidences of incomplete development of the distal limb bud. In other instances, the arm may be fairly well developed with a poorly developed forearm and limb bud for the hand. Usually these small processes are valueless from a functional standpoint and are in the way as far as the fitting of prostheses is concerned

The deformities of the fingers may be manifested in many ways. There may be complete absence of a finger or an entire ray or of the entire radial or ulnar side of the hand. The fingers may be present but incompletely developed, or there may be other associated anomalies such as syndactylism, partial or complete, or symphalangism or any group of such associated deformities.

Treatment

Unfortunately, when there is an absence of a part there is no way to add on or replace the part Usually one must accept deformity particularly if there is incomplete loss of a part such as a forearm arm or hand. In such instances when the child reaches the age of four to five years a suitable functional upper extremity prosthesis should be added. Most children adapt well and tend to utilize prostheses with very little training. If there are small useless appendages present they should be sacrificed in order to gain an accurate fitting of the prosthesis. Children do well with quite short arm and forearm stumps It is amazing to watch the function of a multiplying action joint with a short stump. People usually count stumps but rarely count fingers Any useless part should be sacrificed. The cosmetics are always improved by such procedures, and quite often

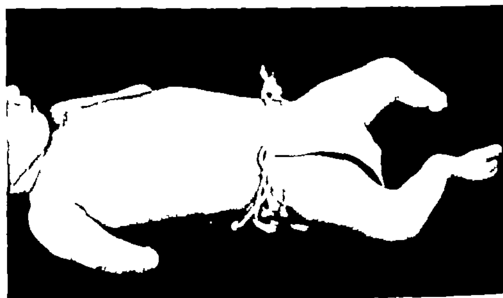


Figure 232 Congenital absence of both forearms and left lower leg with flexion contracture of both knees. Flexion contractures were corrected by posterior surgical release and skeletal traction. The right leg was then braced and a prosthesis was fitted on the left. Patient now walks with crutches at the age of three years.



Figure 233 Congenital absence of the first metacarpal with poorly developed phalanges. Has some function in the long flexor and extensor. Metacarpal graft has recently been performed with the thumb in the opposing position. The final result is not available as yet.

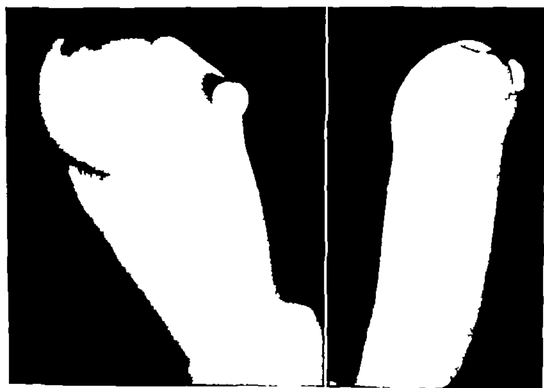


Figure 234 Congenital absence of the hand. Should have a prosthesis fitted at an early age (about four years). *Figure 235* Congenital absence of the forearm and hand. This patient should have a prosthesis at the age of about four years with an elbow joint.

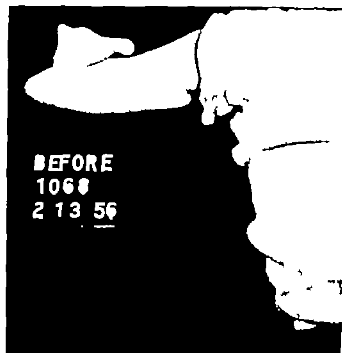
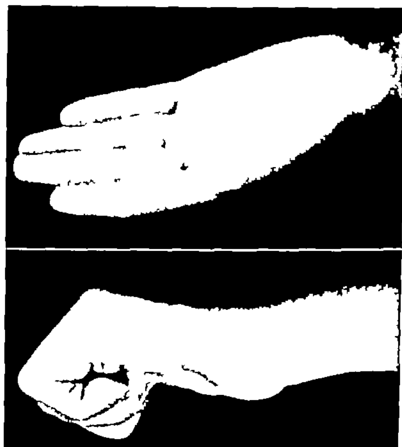


Figure 236 Rudimentary bud of forearm and hand. Better functional and cosmetic result may be attained by sacrifice of the bud and fitting the remaining arm stump with a prosthesis.



Figures 237, 238 Congenital absence of the thumb. Excellent function of fingers



Figure 239 Same case as Figures 237 238. Use of pencil showing substitution pattern. Should not consider surgery unless bilateral then one might transpose the index finger for the thumb on one side

function is also improved. When these children have been born without certain functions they tend to adapt to this loss without too much difficulty, become quite agile and substitute very nicely for the missing parts. This should certainly be encouraged. If phalanges are absent but metacarpals are present, the webs may be deepened, and the metacarpals then used for grasping This is particularly valuable as regards the first metacarpal If the entire thumb ray is absent bilaterally, a pollicization may be performed on one side. It is better to perform this type of surgery in later childhood after the age of five or six years. If syndactylism is present this should be released as described under this heading.

SYMPHALANGISM

This condition results in failure of development of the interphalangeal joints and occurs most frequently as a loss of the distal interphalangeal joint. The condition is frequently hereditary There is no treatment and as a rule the patient does not realize the presence of the deformity and usually adapts well from a functional standpoint.

TRIPHALANGEAL THUMB

Sometimes a thumb develops to appear as a finger This may occur actually with failure of development of the thenar muscles and lack of a wide intermetacarpal space between the first and second metacarpal along with three phalanges in the ray Another variation is a more or less normally developed thumb with three full phalanges This is more satisfactory in that the function is more normal and treatment is usually unnecessary On occasion three phalanges may be



Figure 240 Congenital maldevelopment. Right hand can be improved cosmetically by removing the poorly developed radial and ulnar digits.



Figures 241 242 Same case as Figure 240 Adaptability at early age Function will improve in this case with use and time



Figure 2,3 Multiple congenital skeletal anomalies Child has adapted well to the use of one finger on the right He can lace shoes and tie bows as quickly as anyone

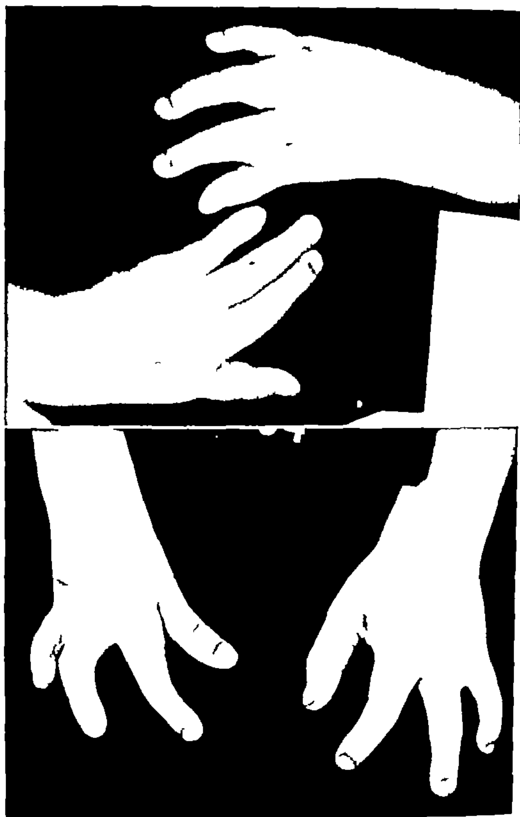


Figure 244 (above) Congenital absence of digit with webbing of the first metacarpal space Child has congenital deafness and congenital anomalies of the ears in addition. *Figure 245 (below)* Same case as Figure 244. Result after Z-plasty deepening of the thumb web.

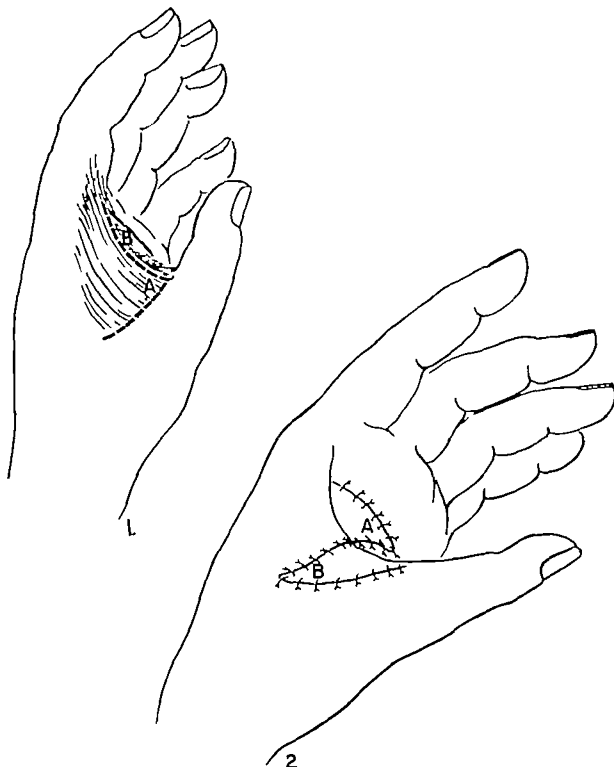
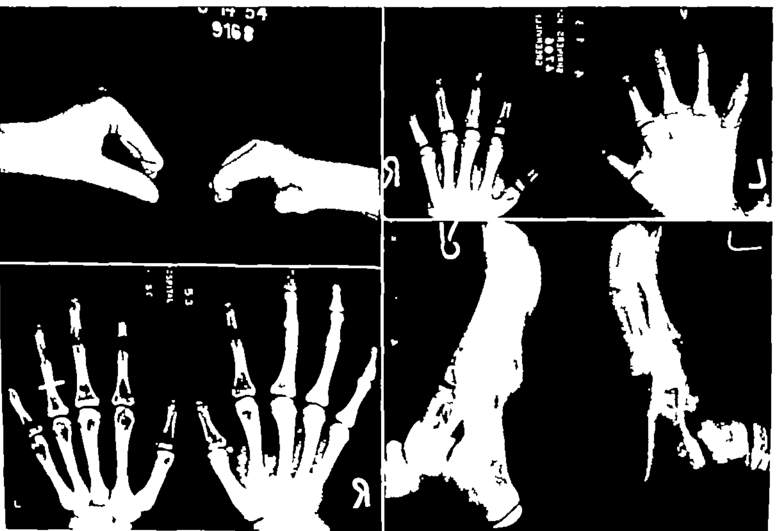


Figure 140 Widening adductor web by Z-plasty to procure additional function of thumb



Figures 247 248 249 250 Symphalangism in mother and daughter X-rays of child's feet show incomplete segmentation of calcis and cuboid and talus and navicular

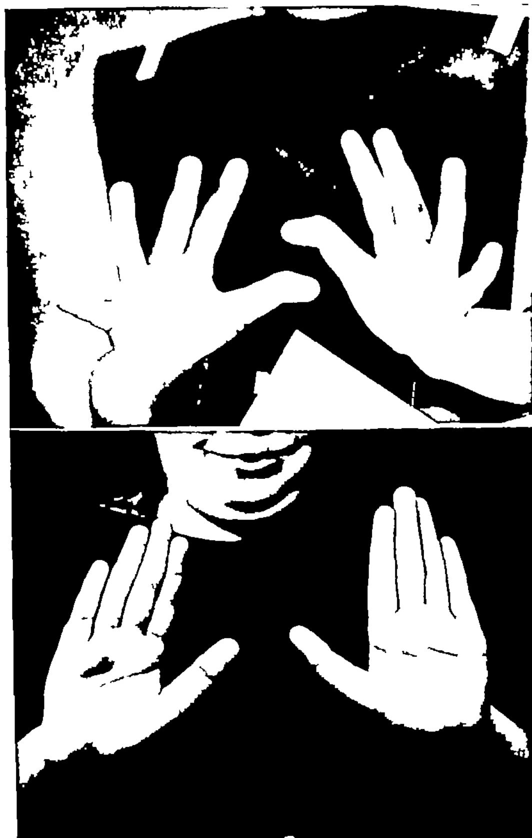


Figure 251 (above) Triphalangeal thumb bilateral, the middle phalanx being wedge shaped. *Figure 252 (below)* Same case as Figure 251 after removal of middle phalanx. Excellent functional and cosmetic result.

present with one of the phalanges deformed or wedge-shaped. In such instances whereby there is a deformity of one of the phalanges and fortunately this is usually the middle, this segment can be removed surgically allowing the other remaining normal phalanges to approximate each other They will form a good joint. The ligaments should be sutured and most often the thumb will have very good function and cosmetic appearance

References

BARSKY A J Congenital anomalies of the hand. J Bone & Joint Surg 33A: 35 1951

BROOKS B AND LEHMAN E. P The bone changes in recklinghausen's neurofibromatosis. Surg Gynec & Obst 38: 587 1924

BUNNELL, STERLING Contractures of the hand from infections and injuries. J Bone & Joint Surg., 14: 27 1932

BURWELL, S Surgery of the Hand. J B Lippincott Co Philadelphia, 1944

COLONNA, P C Some common congenital deformities and their orthopedic treatment. New York State J Med 28: 713 1928.

JOHNSON H. M Congenital cicatrizing bands, report of a case with etiologic observations. Am J of Surg 52: 498, 1941

KANAVEL, A. B Congenital malformations of the hand Arch. Surg 25: 1932.

McCARROLL, H R. Clinical manifestations of congenital neurofibromatosis. J Bone & Joint Surg., 32A: 601 1950

MACCOLLUM D W Webbed fingers. Surg Gynec. & Obst., 71: 782 1940

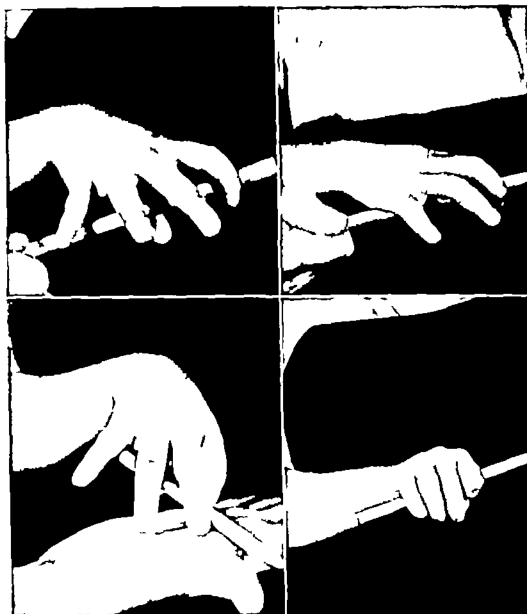
MOORE, B. H. Some orthopedic relationships of neurofibromatosis. J Bone & Joint Surg., 23: 109 1941

SACHS, M D Familial brachyphalangiae Radiology 35: 622 1940

CEREBRAL PALSY

Reconstruction of the hand in cerebral palsy is one of the most difficult tasks confronting the surgeon treating hand conditions. The results are frequently disheartening and in general treatment should be conservative There are very few cases in which surgical intervention is advisable or beneficial. Many factors serve to complicate the treatment of these hands. One of the first and most important steps is the careful classification of these cases into one of the basic types i e true spastics athetoids tension athetoids, ataxias or rigidities. Therapy which may prove valuable in one of these groups can well be completely ineffective or contraindicated in another The classification may at times be difficult making careful observation and repeated examinations necessary before a definite therapeutic plan can be developed.

A second major factor which plays an important part in deciding treatment is mentality Many of these children are retarded and lacking in cooperativeness which is so necessary in hand surgery One of the great factors contributing to the difficulties of treatment is that even in specific groups there are great variations in the severity of involvement. In certain cases, spasticity and poor coordination may seem to play the greatest part in dysfunction, however there may also be an additional definite weakness of the opposing muscle. In the very young cerebral palsied child, major emphasis should be placed on preventing fixed position. The fingers thumb wrist and forearm should be stretched passively several times daily Frequently these children do not recognize the effective hand as such and make no attempt to use it as a hand even in unilateral cases.



Figures 253 254 255 256 These figures show typical attempt of the spastic to grasp Active flexion of the wrist is performed in order to release the spastic finger flexion Incoordinate flexing of the fingers and final grasp through extension of the wrist which enhances the use of the spastic finger flexors Child with this pattern can be trained and surgery should often be avoided.

Early recognition of the hand should be taught using toys, colored beads and blocks to stimulate interest. Occasionally the doctor or parent will attempt to restrict the good hand in order to force the child to use the affected one. This procedure is condemned as it has been definitely shown that restricting a dominant hand may lead to serious psychological difficulties such as stuttering, loss or lack of speech and behavior problems Occasionally after discovering the dominant hand restriction of the other may cure a psychological complex.

SPLINTING AND PHYSICAL THERAPY

Following a period of observation, some of these children may be helped by the use of a small cock up splint or a splint with an outrigger to hold the thumb in extension may be necessary Occasionally rigid splints or plaster bivalves may be used at night or on a part time basis during the day in order to hold the forearm in a supinated position and to aid in passive stretching exercises. Later as the child begins to develop and become more cooperative, active exercises can be added to his treatment program. Physical therapists are helpful in teaching the child and parents specific exercises as outlined by the doctor Major functions to be stressed are those of reaching releasing the fingers stabilizing the wrist in extension, grasping and releasing the fingers Cases with true spasticity usually tend to flex the wrist as the fingers are extended incoordinately They also have difficulty extending the thumb out of the palm. As grasp is attempted varying degrees of wrist flexion are maintained which hinders active grasping function of the fingers Frequently the thumb will be in a position of adduction prior to finger flexion interfering severely with pick up and attempted pinch. In most of these cases if one extends the wrist, thereby aiding the strength of grasp the patient may be unable to extend the fingers effectively for function. Treatment must be carried out many times daily, and parental cooperation and understanding are essential.

The preceding conservative program is used in all types of cases and should be the only type of treatment for ataxias athetoids and tension athetoids In athetoids one of the prime initial efforts should be toward active relaxation on the part of the patient Time is a great factor here but again understanding on the part of the family and helpfulness of those charged with the care of the patient may make a great difference in these cases. It is amazing to see one of these patients attempt to write or use a typewriter while fighting the marked overflow Surgery in some of these cases will produce much poorer function and in very few instances will it produce the desired effect.

Surgical Treatment

Occasionally surgical intervention will definitely improve the function in these hands. At the North Carolina Cerebral Palsy Hospital Goldner found only twelve cases acceptable for hand surgery out of 300 selected as eligible for general treatment taking into consideration their physical status and mental condition.

Prior to selecting a case for surgery it is advisable that the surgeon follow the patient for a long period to make sure that the proposed procedure at least has a

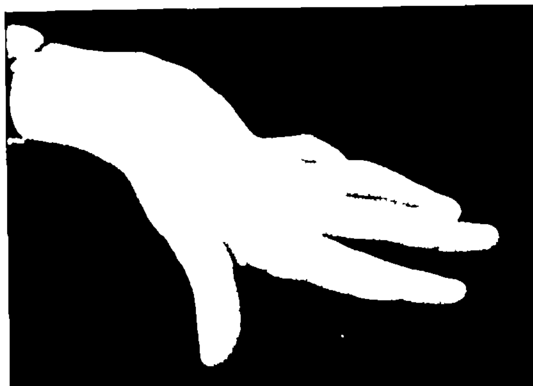


Figure 257 Cerebral palsy spastic type showing reach and poor ability to get the thumb away from the palm

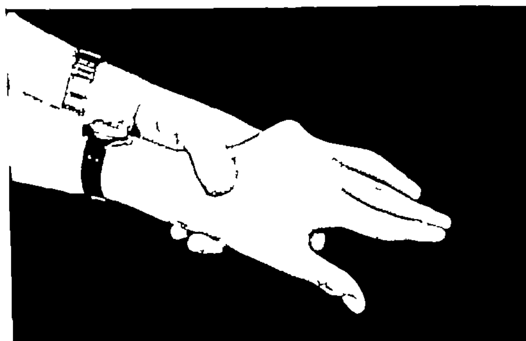
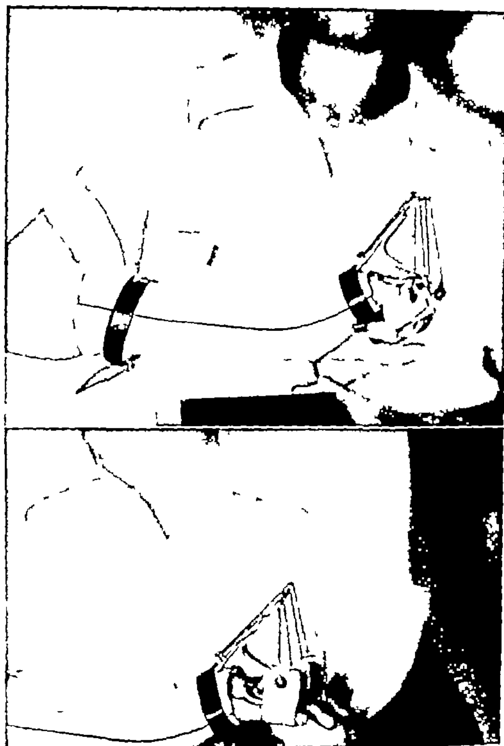


Figure 258 Same case as Figure 257 Patient with wrist immobilized passively in extension and ulnar deviation showing active ability now to get the thumb away from the palm for better function Ability to extend fingers not quite as good but would still be quite effective for reach This case would profit from a wrist fusion.



Figures 259, 260. Removable plastic splint with active rubber band traction outriggers for finger and thumb extension. Bar prevents metacarpophalangeal joint hyperextension. This is used part time in order to attempt to train proper patterns.

reasonable chance of improving function. The use of casts or splints may be extremely helpful in making a decision. The brace or cast may be placed on the extremity, holding it in the position that might be attained by surgery The use of the hand is observed over a period of time in the cast or splint before making a decision. Before proceeding with definitive surgical procedures, it is essential that a detailed examination of the muscles be made Phelps has emphasized that in addition to the charting and evaluation of spastic muscles, the power of other associated muscles must be evaluated and their degree of control noted. The importance of evaluating the power of nonspastic muscles is emphasized because certain antagonistic muscles may show a great weakness and at times may be completely flaccid This is particularly true in the muscles controlling the wrist and fingers If there is a flaccid paralysis in the antagonistic muscles, relieving the spasticity will not solve the problem. Surgery should be used only as an adjunct to the general treatment of the hand in cerebral palsy Many of the operative procedures have been devised for other types of neurological involvement, and it should be emphasized that if these procedures are used on the cerebral palsied child the final results are never as satisfactory The procedures to be used in cerebral palsy for improvement of the spastic hand should be those which stabilize the wrist in order to permit better finger function, to get the thumb away from the palm in order that the hand can be opened and closed effectively and improvement of supination Certain minor procedures may be helpful, and occasionally an excellent result may be obtained Intricate procedures which are often used for polio should be left to the surgeon who is well versed in hand surgery Arthrodesis of the wrist is performed in order to improve the function of the digits and correct deformity of the wrist. The flexor carpi ulnaris is often a strong spastic muscle capable of producing considerable deformity Green found that supination was improved by transplanting this muscle around the dorsal aspect of the forearm to the distal end of the radius Hands requiring stability of the wrist will be helped considerably by arthrodesis just past the neutral position in extension. This position will provide for more effective and efficient use of the fingers. Not infrequently this positioning of the wrist will result in the fingers being tightly clenched in the palm without the ability to extend This condition should be known prior to surgery and a first procedure would be that of transplanting one of the strong wrist flexors into the finger extensors Occasionally this procedure may be all that is necessary however a wrist fusion is usually necessary to provide for adequate stability The thumb in palm deformity is one of the most disabling encountered in the spastic hand. If the deformity is mild ulnar deviation of the hand at the wrist will increase the tension of the extensor and abductor mechanism of the thumb, thereby pulling the thumb phalanges and the first metacarpal toward the radial side. This was first emphasized by Cooper and it is wise in arthrodeses of the wrist to place the hand in moderate ulnar deviation in order that this mechanism may become effective in pulling the thumb up out of the palm. It should be remembered that if the flexor pollicis longus is spastic or contracted and the thumb web is tight no amount of ulnar deviation of the hand will produce an effective result in getting the thumb out of the palm A hypermobile metacarpaphalangeal joint will often result in the de-

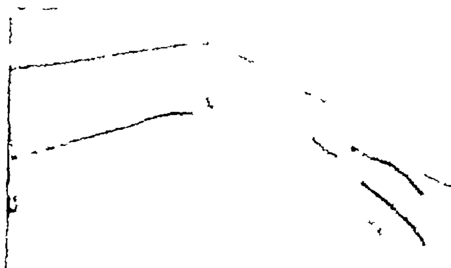
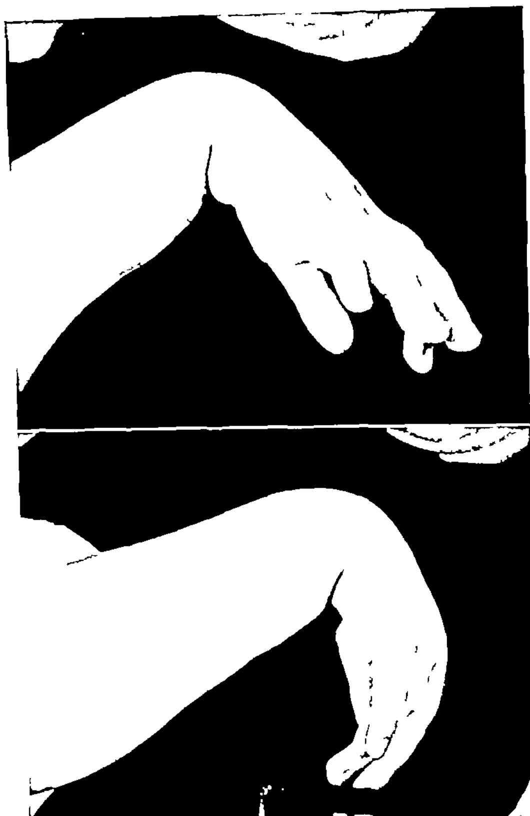


Figure 201 Cerebral palsy showing attempt to extend the fingers and wrist



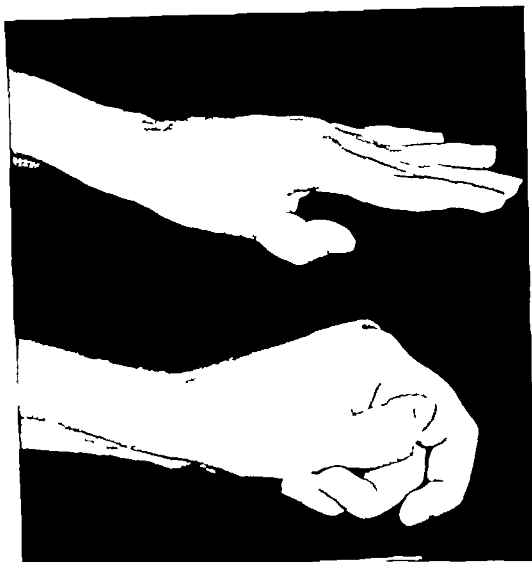
Figure 202 Same case as Figure 201 after transplantation of the flexor ulnaris to the common extensors and the flexor radialis to the extensor pollicis brevis. Patient has a good grip but now hyperextends the wrist and is unable to extend the fingers away from the palm Would be improved now by a wrist fusion in a neutral position. This pattern sometimes occurs after transplantation of wrist flexors into finger extensors due to active insufficiency thereafter of wrist flexion



Figures 263–264. True spastic cerebral palsy showing patient's attempt to extend the fingers and wrist (above) and to grasp (below).



Figures 265 266 Same case as Figures 263 264, showing reach and grasp after wrist fusion in a neutral position.



Figures 267 268 Reach and grasp following transplantation of the flexor ulnaris to the common extensors and the flexor radialis to the extensor pollicis brevis. Prior to surgery the thumb was held flexed and the fingers clenched with the wrist markedly flexed. Now patient has better reach and grasp but the thumb hyperextends This has now been improved by an arthrodesis of the metacarpophalangeal joint.

formity of the thumb in the palm and weakness of the intrinsic musculature of the thumb may lead to the thumb in palm position. Spasticity or contracture of the thumb web may be treated by sectioning the web fascia and stripping the dorsal interosseous muscle from the first metacarpal as well as tenotomizing the adductor pollicis These procedures are often done in conjunction with metacarpophalangeal joint fusion If the extensor mechanism is inactive or weak, transplantation of an active wrist or finger flexor to the extensor pollicis longus should be carried out. Sometimes this leads to an imbalance with hyperextension of the metacarpophalangeal joint and a fusion of this joint should be added



Figures 269 270 These figures show a hand of a cerebral palsy true spastic type following a wrist fusion in ulnar deviation with the wrist in a neutral position regarding flexion and extension These show patient's ability to extend the fingers and to get the thumb away from the palm for grasping Prior to surgery patient clenched the fingers and thumb into the palm and had difficulty attempting to get the fingers or thumb away from the palm.

References

BURMAN M S The spastic hand J Bone & Joint Surg **20**: 133 1938

COOPER, WILLIAM Surgery of the upper extremity in spastic paralysis. Quart. Rev Pediat., 7: 64 1951

GOLDNER J L Reconstructive surgery of the hand in cerebral palsy and spastic paralysis resulting from injury to the spinal cord. J Bone & Joint Surg., 37A: 1141 1955

GREEN W T Transplantation of flexor carpi ulnaris for pronation-flexion deformity of the wrist. Surg. Gynec & Obst., 75: 337 1942.

GREEN W T AND McDERMOTT L J Opreative treatment of cerebral palsy of spastic type J A M A 118: 434, 1942.

McCARROLL, H R Surgical treatment of spastic paralysis, instructional course lectures of the American academy of orthopedic surgeons. 6: 1949 J W Edwards, Ann Arbor Mich 1949

POLIOMYELITIS

Poliomyelitis frequently involves the upper extremity resulting in residual weakness and a poorly functioning hand In some instances, the hand is so severely involved that very little functional value is maintained, however, by certain operative procedures, the value can be increased These consist of rerouting of tendons, thereby preventing or correcting deformities caused by strong musculature and increasing the efficiency of poorly functioning muscle. Occasionally, a completely paralyzed muscle can be replaced by tendon transplantation In other instances, substitution patterns are developed or can be developed with help so that a fairly valuable hand is obtained Occasionally these substitution patterns may be strengthened or enhanced by surgical procedures.

From the very onset of the acute disease the hand should be cared for and treated with function as the final goal The joints should be kept mobile and secondary contractures and deformities prevented by proper utilization of active and passive exercises and splinting in the position of function. During the sensitive stages of the disease, passive splints may be used to rest the parts, preventing stretching of painful, weakened muscles Later as sensitivity and muscle spasm disappear and muscle power begins to return certain active splints are used. These splints are devised to afford helpful stabilization of the wrist action in some 10 or 15 degrees of dorsiflexion. They are made of spring type metal to afford support and yet maintain flexibility Outriggers are used to help with finger extension or flexion, or both, by the use of spring metal or elastic bands. Opponens splints are usually rigid or semi rigid keeping the thumb in the opposed, functioning position They may be made of plastic or metal

In the younger age groups (up to 10 years) it is best to utilize functional splinting for a period of years even though certain operative procedures might be available to improve function thereby replacing the splint. There are several reasons for this recommendation (1) operative procedures are technically more difficult in the young child, (2) coordination for refined hand movements to take over tendon transplants is lacking because of incomplete development of the nervous system, (3) better results are attained with full cooperation which is often lacking in the younger age group (4) bone growth should be as complete as possible ot prevent disproportion between bony framework and the tendon transplantation

There are definite exceptions to waiting, and these exceptions are mainly when the tendon to be transplanted is producing bone or joint deformity in spite of splinting and exercises

There are certain conditions present in polio not always present in traumatic hands which are favorable both to the patient and the surgeon. On the other hand other conditions exist which make rehabilitation more difficult and uncertain.

Unfavorable

(1) Disseminated muscle weakness
(2) Substitution patterns
(3) Involvement of the trunk and other extremities

Favorable
(1) Good skin and subcutaneous tissue
(2) Normal sensation
(3) Good mobility of joints

The procedures devised to surgically improve hand function may be divided into three groups (1) arthrodesis (2) tendon transfers (3) tenodesis. Of these three groups, the tendon transfers are the most valuable and produce a better functioning hand

ARTHRODESES

Arthrodeses should be used only as a last resort. Occasionally, an arthrodesis is indicated to free tendons for other usage but in polio, the loss of joint motion in the upper extremity may be disabling. All other solutions to improve function should have been carefully considered before doing such procedures as fusing the wrist. This procedure should be regarded as a last resort measure, and the relationship of a stiff wrist to the total function of the patient's extremity must be determined. Occasionally it may be seen that the finger and thumb function could *undoubtedly be improved by utilizing the wrist tendons following* stabilization however after careful consideration it may be found that wrist motion will be much more useful than the improvement of finger and thumb function Fusion of the wrist in the wheelchair patient may cause loss of the ability to use the chair thereby disabling him completely because he has lost his means of locomotion In other patients a flexible wrist is necessary in order to shift the body weight. A fusion of the wrist here might well prevent the patient from being able to move about. It is important in all instances to consider body function as a whole in relation to the affected hand before permanently stabilizing a joint. Arthrodesis of the metacarpophalangeal joint is certainly of value and necessary sometimes to correct deformity and to increase usefulness of the fingers however if at all possible, it is to be avoided The avoidance of this procedure is especially necessary in a severely handicapped person who requires a flat hand in order to push himself about. It has also been

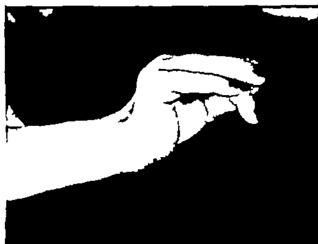


Figure 271 Showing hand in which all wrist flexors have been transplanted into finger and thumb extensors. The stabilizing effect of the wrist flexors has been lost and wrist cocks back and fingers do not *extend in the metacarpophalangeal* joints.



Figure 272 Same case as Figure 271 If wrist is stabilized toward flexion the fingers then extend beautifully It is always best to leave at least one wrist flexor in tact when transferring tendon whether it be for cerebral palsy polio or brachial palsy

found that grasp cannot be too strong thereby making the use of crutches ex tremely difficult. It is wise in the consideration of arthrodesis to wait as long as possible, preferably until maturity The reason for this is that the person should be allowed to seek and try a vocation first. The relationship of the hand to his vocation is extremely important and he may be greatly handicapped by such a permanent procedure as an arthrodesis.

The bone block procedure may be helpful in improving the function of the hand, but it should be remembered that it is a poor procedure of the patient is a crutch walker

Arthrodesis of the interphalangeal joint of the thumb is valuable if other means of stabilization are found impossible.

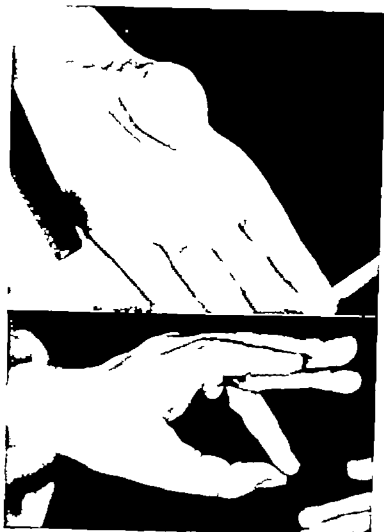
TENDON TRANSPLANTATION

It has been found that tendon transplants are by for the best procedures to increase the function of the hand. Occasionally, active motors are not available and it has been found best to perform passive or active tenodeses which will be discussed later Moving a tendon must never be done without careful considera tion and an accurate muscle evaluation It is frequently necessary to retest the muscles several times in order to get an accurate picture of the muscle power Frequently it is difficult in the young child to obtain an accurate muscle test. It is to be remembered that occasionally two muscles acting together may produce a functioning unit, and yet each muscle will rate only fifty per cent of normal. A muscle used for transplantation should rate at least eighty five per cent of normal

It should be remembered that transplanting a tendon should not excessively weaken that portion of the hand from which it is taken. A good example of this is the transplantation of a strong wrist extensor to gain finger flexion or opposition when the remaining wrist flexors are weak. This will result in the loss of stabilizing function of wrist extension and thereby defeat the purpose of the original transplant to strengthen the grip If there is any doubt regarding the



Figure 273 A Loss of opponens Atrophy of thenar eminence.



Figures 274, 275 Same case as Figure 273 after opponens transplantation using the sublimis of the ring finger transplanted to the base of the proximal phalanx of the thumb, using the flexor carpi ulnaris as a pulley

power of a muscle, an incision should be made over the belly, and the color and texture of the muscle should be inspected

In adults, it has been found advisable to wait for a period of two years prior to any tendon transplantation In children up to the age of twelve, it is best to wait for even longer periods Proper splinting and substitution patterns may evolve which will make surgery entirely unnecessary In children, it is better to err on the side of waiting too long than to perform surgical procedures without careful consideration.

Opposition

Paralysis of opposition is a major disability The transplant of choice for restoring opponens function is to use the sublimis tendon of the ring finger as a motor The prerequisites for this procedure are (1) the profundus of the ring finger must be adequate for flexion of the digit without the sublimis, (2) a strong flexor pollicis longus (3) a strong extensor pollicis longus and abductor pollicis longus, (4) a good or normal flexor digitorum sublimis, (5) the absence of bone, joint or soft tissue deformity of the thumb, (6) a strong flexor carpi ulnaris and (7) functional strength of the fingers and the palmar arch.

If these conditions do not exist the profundus of the ring finger may be reenforced with an adjacent profundus or sublimis The flexor longus, abductor or extensor of the thumb may be strengthened by another transplant. Of these the most suitable tendon for this is the long extensor of the wrist. The indicis proprius can often be used to great advantage for strengthening the extensor pollicis longus The flexor carpi ulnaris may be reenforced by the extensor carpi ulnaris. These procedures will be described in detail later If there is deformity or contraction of the thumb, it should be corrected prior to any attempts at tendon reconstruction Sometimes a tenotomy of the adductor of the thumb is necessary to release soft tissue contracture of the web space Permanent tightness of the joints may be relieved by a rotation osteotomy of the first metacarpal Occasionally in a thumb with a strong flexor pollicis brevis and no disability other than a weakened or absent opponens an osteotomy of the first metacarpal may produce a satisfactorily functioning result obviating the necessity of an opponens transplant.

The opponens transfer should pull from the region of the pisiform for best mechanical efficiency When using the sublimis this is accomplished best by bringing the tendon beneath and around the ulnar side of the flexor carpi ulnaris at its insertion as a pulley The tendon should be attached to the thumb on the ulnar side of the proximal phalanx. The transplanted tendon should pass across the base of the palm subcutaneously from the pisiform to the metacarpophalangeal joint of the thumb Proper tension is of utmost importance and is gained only by experience with the procedure If it is inserted too loosely, it will lack sufficient strength to properly oppose If it is inserted too tightly the tendon may migrate over the metacarpophalangeal joint and cause a hyperextension contracture Following transfer the hand should be immobilized in a plaster splint with the wrist in approximately 30 degrees of flexion and 15 degrees of ulnar deviation Immobilization should be continued for a period of three weeks

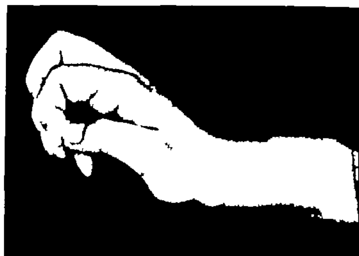


Figure 276 Typical pinch of opponens loss plus a weak first interosseous.

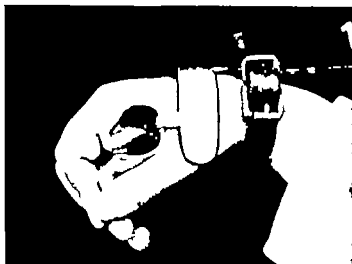


Figure 277 Same case as Figure 276, showing the support given by an opponens splint.

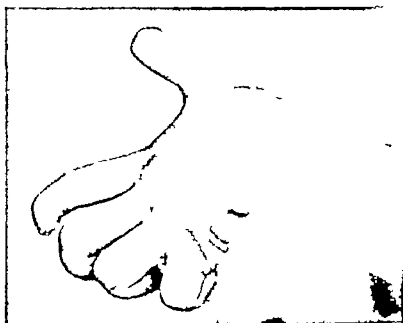


Figure 278 Effect of imbalance of the long flexors, intrinsics and long extensors due to weak wrist extensors. Thenar muscles are absent except for the long thumb flexor and extensor and long abductor

followed by active exercises, using an opponens splint for protection between exercise periods If the sublimis of the ring finger is not sufficiently strong, the sublimis of the middle finger may be used satisfactorily Occasionally, it may be necessary to reenforce a transplanted weaker sublimis. This can be done by suturing an additional tendon into the musculotendinous junction of the sublimis tendon. In the absence of sublimi, the extensor carpis ulnaris may be used, prolonging its length with a free tendon graft. It is passed around the ulnar border of the wrist proximal to the dorsal carpal ligament thereby requiring no pulleys.

Rerouting of the Extensor Pollicis Longus

Frequently, the thumb will drift into ulnar deviation at the metacarpophalangeal joint. This position will often not be corrected by opponens transfer This condition may be alleviated and the opponens transfer made more effective by rerouting the extensor pollicis longus tendon.

Procedure The dorsal carpal ligament is sectioned, and the extensor pollicis longus removed from its groove and brought volarward in the subcutaneous tissue Its position is maintained by a subcutaneous suture so as to hold the tendon in direct line with the thumb ray The dorsal carpal ligament is then resutured.

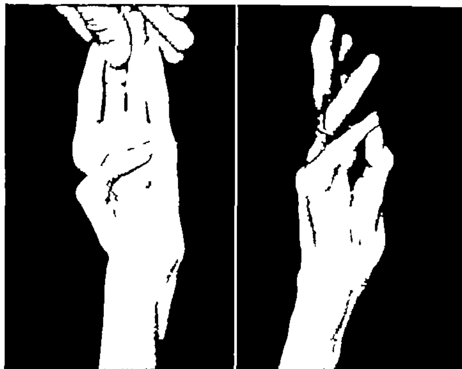
Reenforcement of the Pulley for Opponens Transfers

At times the opponens transfer will migrate toward the radial border of the wrist, thereby reducing the power and effectiveness of the transplant. The usual reason for this migration is weakness of the flexor carpi ulnaris. This procedure can be corrected by suturing the extensor carpi ulnaris into the tendon of the flexor carpi ulnaris

Procedure A hockey stick incision is made over the volar ulnar border of the wrist. The flexor carpi ulnaris is then freed and the forearm is pronated The extensor carpi ulnaris is isolated and sutured through a slit in the flexor carpi ulnaris Fixation is maintained by interrupted silk sutures The anastomosis is performed just proximal to the route of the opponens transfer around the flexor carpi ulnaris tendon.

The index finger stands next to the thumb in functional importance Loss of intrinsic power of this finger along with weak or absent opposition of the thumb is more commonly seen in poliomyelitis than weakness in all the remaining extrinsic muscles of the hand Absent intrinsic power of the index finger prevents extension of the distal two phalanges and the inability to abduct the finger into a position opposite the opposed thumb in the formation of the pinching mechanism Strong extrinsic flexors and extensors of the index finger do not replace the dorsal and volar interossei and lumbricale muscles in the pinching mechanism The clinical picture is usually that of cocking or hyperextension of the metacarpophalangeal joint when the function of pinching is attempted. This is due to a loss of stability in flexion of the metacarpophalangeal joint.

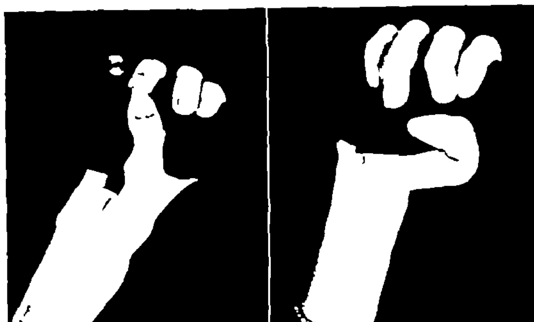
In attempting to restore the intrinsic factor particularly with regard to stability in flexion of the metacarpophalangeal joint transfers must insert into



Figures 279 280 Opponens transplant functioning well but there is practically no abductor brevis or flexor pollicis longus to give stability

the lateral band and function is better mechanically when the insertion is from the volar aspect of the hand Many procedures have been described for the restitution of this function Probably the most popular is that of sublimis transfer into the lateral band as described by Bunnell. Even in the absence of active functioning musculature tenodeses are sometimes performed to stabilise these joints according to the methods of Fowler and Riordan. These procedures will be discussed to some extent later under Tenodeses Frequently the indicis proprius is utilized transferring it into the lateral band tendon to afford good abduction of the index finger This transplant restores abduction and extension but lacks stabilization of the metacarpophalangeal joint in flexion. Irwin and Eyler have described a procedure which is excellent for the restoration of the abduction element in flexion and extension and also the stabilizing effect of intrinsic power on the metacarpophalangeal joint. This is the split sublimis procedure.

Procedure A long finger sublimis is severed at the proximal joint of the finger and withdrawn at the musculotendinous junction. The tendon is split into two tails. One tail is introduced through the lumbricale canal and inserted into the lateral band past the lumbricale insertion. The other is inserted into the volar and dorsal interosseous tendons of the first dorsal interosseous. The wrist is kept in neutral position and the metacarpophalangeal joints flexed 80 degrees with the interphalangeal joints fully extended Immobilization is for a period of three weeks following which active exercises are started.



Figures 251 252 As strength is added, the grip is lost and the thumb flexes. This can be avoided by inserting one slip of the sublimis into the metacarpal and one into the phalanx so that the metacarpal is brought into opposition equally with the rest of the thumb

Finger Flexion

Irwin is of the opinion that weakened profundi may be successfully reenforced in continuity with other strong profundi or sublimi. When this procedure is performed properly excellent results have been obtained. The procedure is accomplished by splicing the strong tendons into the weakened ones proximal to the volar carpal ligament. The wrist extensors work more or less synergistically with the long flexors of the fingers and it has been found that wrist extensors work well in restoring finger flexion. The extensor carpi radialis longus has a good excursion and works well when it is woven into the profundus tendons at the musculotendinous junction The anastomosis should be made under considerable tension with the little finger being under the greatest amount of tension and each adjacent finger being placed under relative amounts of tension. The anastomosis should be made with the fingers in a position of relaxed flexion. A plaster splint is applied and removed in about three weeks followed by active motion Prior to utilization of an extensor of the wrist as a transplant particularly to the flexor side the ability to extend the wrist must be assured in the absence of this tendon. This can be guarded against by directly inspecting the remaining wrist extensor muscles. If a strong extensor carpi radialis tendon is not available the extensor carpi ulnaris may be transferred into the flexor digitorum profundus tendons with good restoration of finger flexion. The excursion of the flexor carpi ulnaris is not great enough to give satisfactory finger function. The brachioradialis will work moderately well but its excursion is less than the long extensor of the wrist. Restoration of common extensor action

will restore the ability to extend the metacarpophalangeal joints and i these joints, but will not extend the interphalangeal joints in the ab good intrinsics. The extensor carpi radialis will also work very well for r common extensor function. Its transfer to the finger extensors is mor effected and with less concern than is necessary in moving the same te the flexor side, inasmuch as it can work as a stabilizer of the wrist whe tomosed into the finger extensors.

Procedure The extensor of the wrist is sectioned at its insertion into t of the metacarpal freed up in a retrograde fashion from under the carp ment and then redirected along the dorsal surface of the forearm subcuta and anastomosed into the extensor tendons at their musculotendinous ju The anastomosis must be made with the extensors in considerable tensi the metacarpophalangeal joints in extension. Relative differences in ten accomplished by suturing the little finger tighter than the adjacent rin and the ring finger slightly tighter than the middle finger, etc The e carpi radialis brevis can also be used to replace extensor communis fi The extensor carpi ulnaris is a fair tendon for this purpose The brachio has been found to be an excellent motor for this purpose The wrist flexor synergistic with finger extension may be used quite satisfactorily as m ments for common extensor function The flexor carpi radialis and ulna be used successfully however they are not as satisfactory as the radia sors of the wrist.

Restoration of Finger Function by Tenodesis

As has been previously discussed an arthrodesis in the hand paraly polio is rarely indicated. The procedure might be considered in order valuable tendons for transplantation to weakened fingers and thumb h in evaluating the activity of the patient, one might find the loss of flexib the wrist to be a great handicap Recently Irwin and his associates at Springs have found instances whereby these cases may be aided consi by the use of a tenodesis. Wrist motion can be retained and whereby t ventional transfers could not be performed because of insufficient musc the tenodesis action can be utilized with quite effective results

The function of grasp with the wrist extended is the result of two t muscle action namely contraction of the flexor muscle mass, producing mary flexion of the fingers. In addition extension of the wrist gives ri secondary flexion of the fingers by increasing the distance between the and insertion of the long flexors Conversely flexion of the wrist may the fingers as a result of increasing the distance from the origin to the in of the long extensors. In the presence of weakness or loss of the long ex or flexors of the fingers this function of grasp may be enhanced greatly b odesing the afflicted motors The fixation can be accomplished by imp the common tendon mass into bone creating a tenodesis or by transferr actively contracting muscle into the tendon mass and forming a dynam odesis Irwin has emphasized that the distinction between dynamic te and a standard transfer is difficult. A dynamic tenodesis is a transfer

when inserted into the common tendon group, such as the flexors of the fingers, relies mainly on wrist action for its function with some reenforcement from the transplanted active muscle These tenodesis procedures should be reserved for those patients who lack sufficient motors for conventional transfers The patient must possess a mobile wrist that he can manipulate into flexion and extension with mobile fingers and a functional elbow An active wrist extensor greatly enhances the value of a flexor tenodesis, however, in the absence of active wrist extension the wrist may be made to fall into the extended position by gravity when the forearm is supinated and the elbow flexed The grasp that results is necessarily very slight, but an effective hook is created that the patient may utilize to considerable advantage Active wrist flexion is less important to the grasp than is wrist extension. In the absence of active motors the wrist normally falls into a position of pronation and flexion when the elbow is flexed The status of the intrinsics of the fingers plays an important role in the final result Extensor tenodesis will not fully extend the fingers unless active intrinsics are present Interestingly enough, finger extension may improve with flexor tenodesis if the lumbricales are functional Lumbricale efficiency is increased when the profundi are stabilized

Technique as described by Irwin Flexor tenodesis is performed through a semilunar incision overlying the volar surface of the wrist. The median nerve is identified and carefully retracted from the field The profundus tendons are identified by applying manual traction to each tendon and observing for flexion of the distal phalanges. The position of the fingers is adjusted to correspond to that assumed by the normal hand lying at rest on a flat surface with the wrist supinated. If the tenodesis is to be of the passive type a small window is made in the distal end of the radius and the profundus tendons inserted into the bone with the aid of a single wire suture. Should a dynamic tenodesis be indicated, the position of the fingers is fixed by anastomosing any available transfer into the common tendon mass. Anastomosis is secured with the aid of twisted steel wire sutures The technique for insertion of the extensor tenodesis is the same The common extensors are isolated through a semilunar incision on the dorsum of the wrist which passes to the ulnar side of the tubercle of Lister Anastomosis is accomplished with the hand in a position of rest. Active transfers are inserted under moderate tension Immediate postoperative immobilization is secured with dorsal and volar plaster splints. Flexor tenodeses are immobilized with the wrist in full flexion and the fingers extended and extensor tenodeses with the wrist in full extension and the fingers extended Three weeks postoperatively, the splints are removed and mobilization of the hand and wrist is initiated

References

BUNNELL, STERLING Surgery of the intrinsic muscles of the hand other than those producing opposition of the thumb J Bone & Joint Surg 24: 1942

BUNNELL, S AND HOWARD L. D Additional elastic hand splints. J Bone & Joint Surg., 32A: 226 1950

BUNNELL, STERLING Active splinting of the hand J Bone & Joint Surg., 28: 732 1946

COONRAD R., EYLER, D AND IRWIN C E. Split sublimis transfer for paralysis of the intrinsic muscles of the index finger Correspondence Club Letter Duke Training Program 1951

EYLER, D L, AND MARKEE, J E. The anatomy and function of the intrinsic musculature of the fingers. J Bone & Joint Surg., 36A: 1 1954

FORRESTER, O Fusion of metacarpals of thumb and index finger Acta chir scandinav 67: 350 1931

GOLDNER, J L. AND IRWIN C E An analysis of paralytic thumb deformities. J Bone & Joint Surg., 32A: 627 1950

IRWIN C E. Apparatus for the upper extremity disabled by poliomyelitis Am. Academy of Orthop Surg., Instructional Course Lectures, 9: 1952

IRWIN C E. Dynamic Tenodesis, presented Am. Academy of Orthop Surg., Jan 1956. 257 1942

IRWIN C E. AND EYLER, D L. Surgical rehabilitation of the hand and forearm disabled by poliomyelitis. J Bone & Joint Surg., 33A: 825 1951

KIRKLIN J W AND THOMAS, C. G Opponens transplant an analysis of methods employed and results obtained. Surg. Gynec & Obst 86: 213 1948

RIORDAN D C Tendon transplantation in median nerve and ulnar nerve paralysis. J Bone & Joint Surg., 35A: 312 1953

ROYLE, N D An operation for paralysis of the intrinsic muscles of the thumb J A.M.A., 111: 612, 1938

THOMPSON C F Fusion of metacarpals of thumb and index finger to maintain functional position of the thumb J Bone & Joint Surg 24: 907 1912

RHEUMATOID ARTHRITIS

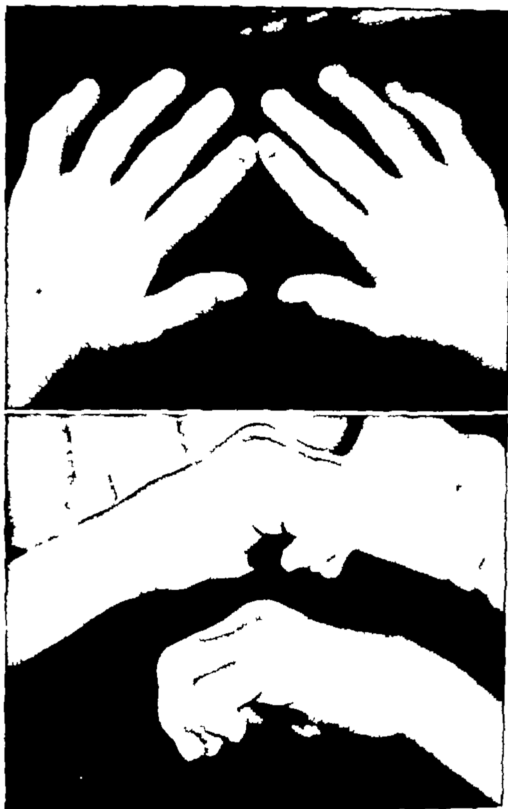
Rheumatoid arthritis is a generalized systemic disease affecting both children and adults In children the disease may vary considerably in its mode of onset. Frequently it begins in one joint and subsequently spreads to become generalized involving all joints Occasionally it will remain localized to one or two joints becoming a problem in diagnosis Its appearance may be manifested by generalized joint involvement with marked toxicity, anemia weight loss splenomegaly and fever

When the hands are involved marked deformity frequently results with severe permanent dysfunction and disability Primary joint involvement is not the only deforming factor Secondary muscle spasm and eventual myostatic contracture contribute to the hand deformity The muscles are in spasm because of irritation of the synovial reaction but later they become fibrosed and permanently shortened Microscopically the muscles tendons and ligaments show multiple foci of inflammation and later fibrotic contracture Some ligaments actually degenerate allowing for subluxation of the joints Some of the tendons may degenerate and rupture at focal spots of granulation The joint changes ligamentous and tendinous involvement all play a part in the deformity but position of the deformity is largely due to a muscle imbalance between the long flexors long extensors and the intrinsics. Because of this disturbance of the normal equilibrium in these three groups of muscles all of the clinical forms of arthritis involving the hands and fingers result in two types of deformity namely the clawhand and the pillroller hand. In the clawhand the metacarpophalangeal joints are pulled into hyperextension by the long extensors thus overcoming the resistance of the intrinsic muscles The fingers flex in the middle joints and extend in the distal joints The more usual deformity of the two is the pillroller hand in which there is flexion of the metacarpophalangeal joints due to the intrinsic contracture The middle and distal joints are thus pulled

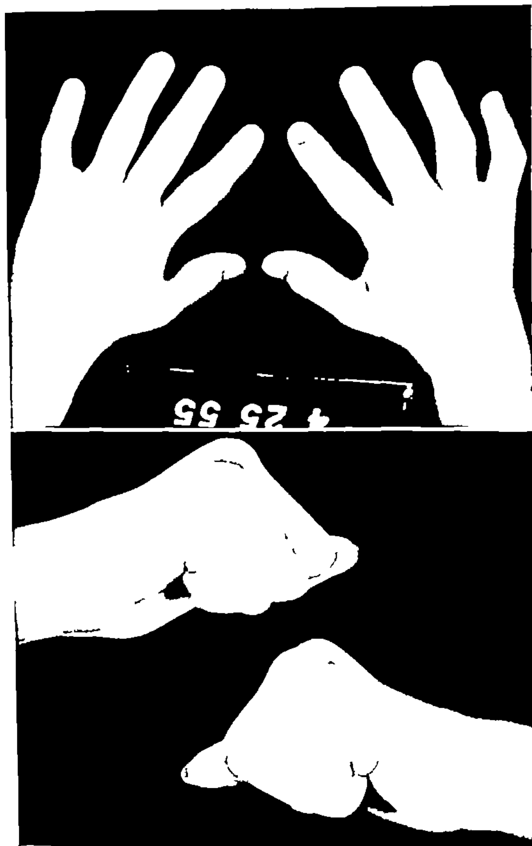
straight by the lateral bands or at times the middle joint may be hyperextended and the distal joints may flex. The position of the wrist seems to play a great part in the effect of the deformity on the hands and fingers in both of these conditions In the claw hand the wrist is in flexion, therefore the passive tension on the extensors leads to hyperextension in the metacarpophalangeal joints In the pillroller hand, the wrist remains in extension and the finger extensors are relaxed The spastic contracture of the intrinsic muscles which are not being opposed by the relaxed finger extensors cause the metacarpophalangeal joints to become markedly flexed The interphalangeal articulations assume a position of hyperextension because of the contracture and tightness of the intrinsic musculature The thumb is usually drawn into flexion and adduction. The imbalance between the short flexors and adductors and the abductor group usually results in adduction and flexion of the carpometacarpal joint The metacarpophalangeal joint is overflexed causing hyperextension of the distal joint because of the pull of the long extensor Deformity of the thumb is more often noted in the pillroller type of hand.

Treatment

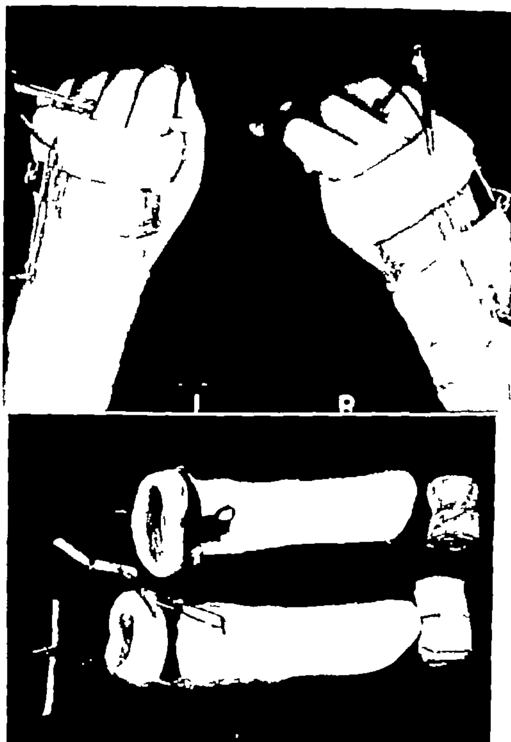
Because of the systemic nature of this disease the treatment of rheumatoid arthritis must first of all be generalized however, great attention should also be given to the local involvement and the prevention and correction of deformity It is rare to see a patient who has in any way been neglected from a general standpoint It is not unusual, however to find that the general treatment has been carried out without emphasis on the local care of joints in an effort to prevent crippling contractures With the advent of newer remedies for rheumatoid arthritis and the improvement in general care and treatment, conservative local measures should become much more efficient and promising in the prevention and correction of deformities such as those that occur in the hand. In those cases seen early prior to the development of contracture the treatment to prevent the deformity should start early and proceed actively under careful observation. The development of deformity is insidious and frequently the doctor and family procrastinate until a serious deformity results. The family physician or pediatrician should handle the general medical care of the patient, but the orthopedist should see the child early during the stage of irritation and painful swelling before the occurrence of permanent contracture and deformity By anticipating the development of deformity due to the imbalance of musculature, proper splinting may be instituted avoiding completely the occurrence of serious disability Drs Bunnell and Steindler have emphasized that the muscles are contracted from irritation and are in a state of spasm It is only later that they become structurally changed by progressive fibrosis Steindler has stated that a spastic muscle is an extensible one which has retained its elasticity while a fibrosed muscle has lost its elasticity The arthritic muscle remains in a reflex spastic state for a considerable period. The correction of this reversible condition by mechanical means and restoration of balance by muscle reeducation offers a wide therapeutic opportunity for the acute and subacute case of rheumatoid arthritis These factors greatly improve the prognosis for the arthritic



Figures 233, 234 Rheumatoid arthritis prior to therapy showing the range of extension and flexion



Figures 285, 286. Same child ten months later after use of paraffin baths, whirlpool and active exercises along with active splinting.



Figures 237, 238 Active splints made of plaster with rubber band traction which can be removed for use and for physical therapy

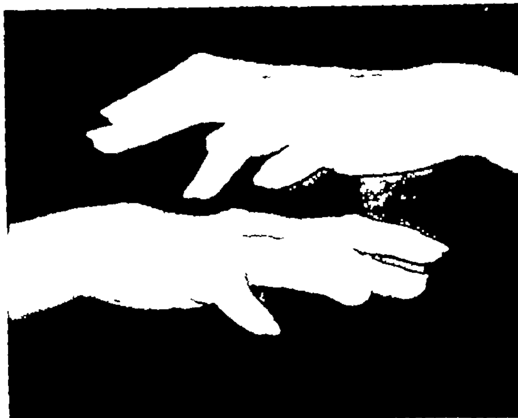


Figure 250 Hands in rheumatoid arthritis with more advanced stage of *deformity show*ing flexed wrist, hyperextended proximal finger joints and flexed middle joints and extende distal joints.

hand in children because they are seen prior to the development of fixed con tracture and bone deformity Even in those instances where definite contrac tures have developed they are usually still in the stage of reflex spasm rathe than a fixed myostatic contracture Physical therapy in the form of heat splint ing paraffin baths and active exercises should be utilized to their fullest Surger is rarely indicated In the early stages of swelling pain and muscle spasm paraffin and whirlpool baths are more effective Development of early flexio contracture of the wrist can be prevented by passive splinting with a cock u splint. Active use of the hand should be encouraged, and fixed immobilizatio is to be avoided. Splints should be of the retention variety designed only t maintain position and should not be applied for corrective effect.

Active splinting is quite effective The active splint should perform one pur pose only however several splints may be used in the same case at differen periods each exerting a different type of effect. Active splints using mild elasti traction or spring effect may be made by the bracemaker If these are no available simple plaster splints using spring wire or elastic traction can b fashioned inexpensively By utilizing the active splints part time to help over come muscle spasm and muscle imbalance fixed deformities can be prevente

Active exercises for the fingers wrist and forearm are essential in the treat ment and should be combined and properly balanced with gentle passive stretch ing of tight joints Emphasis is placed on gentle stretching and manipulativ

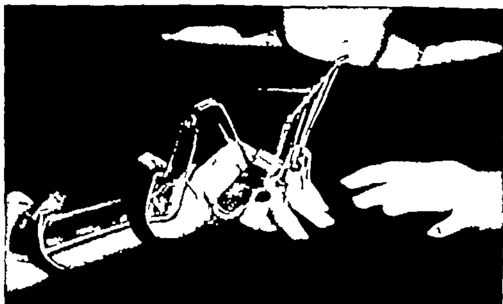


Figure 290 Metal active splint to hold wrist in dorsiflexion while bar prevents metacarpophalangeal extension during active stretch on the proximal interphalangeal joints. This splint is worn part time while gentle passive stretching and active exercises are used at other times along with active use of the hand in general daily activity

treatment, being careful never to approach the point of tearing capsular tissues. The treatment program is attended by some increase in pain which may be allayed by the administration of salicylates Children usually tolerate treatment better than adults and quite cooperative.

The utilization of the foregoing measures accompanied by proper education of the family regarding the chronicity of the treatment will usually produce gratifying results. Periods varying from two to ten years are usually necessary for the disease to become completely burned out and some residual deformities may be expected at the end of this period, however, major crippling can usually be minimized and in many instances it can be completely prevented

References

BUNNELL, STERLING Surgery of the rheumatic hand. J Bone & Joint Surg 37A: 59 1955

BUNNELL, S AND HOWARD L. D Additional elastic hand splints. J Bone & Joint Surg 32A: 226 1950

BUNNELL, S Active splinting of the hand J Bone & Joint Surg., 28: 732 1946

PEACOCK, EARL, JR. Dynamic splinting for prevention of deformities and correction of hand deformities. J Bone & Joint Surg., 34A: 789 1952

STEINDLER, ARTHUR Arthritic deformities of the wrist and fingers. J Bone & Joint Surg 33A: 849 1951

TRAUMA

Trauma in its many varieties accounts for a vast number of hand disabilities and deformities. This is due to increased mechanization of the modern age. Children are susceptible to many of the same types of trauma as are adults, and their reaction to the injury is practically the same with minor variations. Most of the variations have to do with skin thickness and bone growth, bone healing and joint mobility Some of these variations from the adult hand tend

to be helpful in treatment while others make the treatment more difficult. Children have surprising powers of adaptation and are able to accommodate well to the loss of a part. They are able to substitute functions and accept their disabilities and deformities better than one might first suppose. Sometimes it is best to delay large reconstructive procedures until later in life. This may result in sufficient accommodation to the difficulty so that nothing further need be done. On the other hand, young children do not cooperate well, and their small extremities are hard to immobilize making the postoperative care difficult and sometimes unsuccessful.

The basic principles of treatment of traumatic deformities in children are the same as those used in adults, therefore, in this section on trauma the entire field will not be covered. There are certain injuries which are common in children. These will be discussed in detail. Variations in basic treatment of other types of trauma will be brought out.

VOLKMANN'S ISCHEMIC CONTRACTURE

Volkmann's ischemic contracture is one of the most serious deformities of the hand and forearm, and the most dreaded complication of injuries about the elbow and forearm particularly in children. Steindler has stated that from a purely pathological viewpoint ischemic contracture is a fibrous reaction of the muscles of the forearm particularly those of the flexor group. The contracture may involve the muscles *in toto* or in part, and there have been cases reported where muscles of the hand alone were involved. In one instance, all of the muscles escaped involvement except for one sublimis muscle.

The fully developed ischemic contracture actually is a combination of deformities. The usual characteristic picture is that of pronation of the forearm, flexion of the wrist and interphalangeal joints and hyperextension of the metacarpophalangeal joints. The hyperextension of the metacarpophalangeal joints is largely a result of passive insufficiency of the extensors and not necessarily indicative of intrinsic loss. This description is the most frequent type that is observed but other variations may be encountered. Common variations are the absence of hyperextension of the metacarpophalangeal joints and a flexion contracture of the wrist similar to that seen in radial nerve palsy. The thumb is frequently involved with an adduction contracture and the inability to oppose placing it in a plane with the involved fingers.

Etiology

Numerous investigators have tried to solve the problem of pathogenesis of this lesion following its original description by Volkmann, and there have been excellent papers written on this subject both from clinical and experimental standpoints. As one reads these descriptions the impression is obtained that each investigator has proven his premise to be correct. Bardenheuer brought out the venous obstruction theory and this has been adequately substantiated by other authorities in the field. Brooks and his associates have done some excellent experimental studies and have proven quite conclusively that the lesion can be produced experimentally by complete venous obstruction. Volkmann's original idea of an arterial deficit has been substantiated by numerous investigators and experimentation.

Griffiths has produced an excellent work giving convincing proof that the lesion is due to arterial occlusion and muscle infarction. There are those that hold that the lesion is merely an unusual variety of contracture due to nerve injury. In many instances it is undoubtedly true that there is peripheral nerve damage in addition to the contractures; however, there has been no definite proof that direct injury to the nerve is the primary cause of ischemic contracture. There can be little doubt, however, that constriction lasting over a long period of time sufficient to close large vessels will not exert pressure over the median and ulnar nerves with subsequent neuritis and degeneration. Leriche and his workers attributed great significance to the irritation of the sympathetic nerve supply resulting in peripheral vasoconstriction.

In view of these conflicting clinical and experimental impressions, the etiology of Volkmann's contracture could be either arterial or venous obstruction. It is also not illogical to assume that in certain cases all of the previously postulated factors might well play a part.

Because of the popularity of circulatory obstruction as the etiology of Volkmann's ischemic contracture, constricting casts, bandages and dressings have long been associated with the condition. There are cases on record in which no dressing or cast was used, and yet the condition developed. In spite of this, casts or bandages can be very definite contributing causes and for this reason, careful supervision and vigilance are essential features in the treatment of all injuries about the elbow and forearm.

Pathology

In the initial stages the subcutaneous tissues become cyanotic and infiltrated with blood. The superficial veins engorge and there is swelling of the extremity. Deep fascia and muscle envelopes are tight and when cut tend to gap and retract markedly showing a very pale yellowish-gray musculature. In later stages, *the muscles may become gray with shortened muscle fibers and evident* fibrous cords. Within forty-eight hours after the onset, polymorphonuclear leukocytes accumulate about the fibers adding a typical inflammatory appearance. Organization of the exudate begins within four or five days and is usually well established within ten days. Destruction of muscle fibers varies directly with the severity of the case. All gradations of involvement can occur, and in mild cases almost all of the muscle fibers function normally, while in severe cases all of the muscle substance may be replaced by connective tissue.

Histologically, the connective tissue forms in bundles which ramify throughout the muscle bellies replacing necrotic muscle fibers. The fibrous tissue has a fibrillar aspect and forms a meshwork in which there are isolated areas of viable muscle tissue.

Clinical Picture

Following the treatment of an acute injury such as a fracture or dislocation, there is usually a period of relative comfort to the patient. The earliest signs and symptoms usually appear within four to six hours but may develop as late as forty-eight hours. There are recorded cases in which the symptoms have developed as late as a week after the initial injury. Following the period of initial



Figure 291 Typical deformity seen in Volkmann's contracture There is atrophy of the forearm acutely flexed wrist and interphalangeal joints with hyperextended metacarpophalangeal joints.



Figure 292 Severe Volkmann's contracture in eight year-old girl—attempted flexion. Pedicle graft over the volar aspect of the lower forearm has been performed at the site of a skin slough.

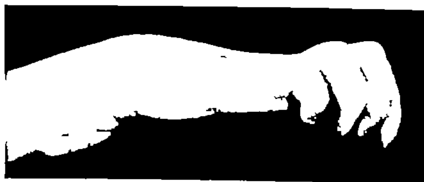


Figure 293 Attempted extension. Only wrist extensors, common extensors and thumb extensors were functioning below the elbow The function of these muscles was greatly inhibited due to the flexion contractures

comfort, there is usually a sudden onset of severe pain in the arm and in the volar aspect of the forearm. The fingers become cold, cyanotic and swollen. The pain increases in intensity, often being incompletely relieved by narcotics. Paresthesias develop, and even though the fingers may feel numb, they are often very hyper-sensitive to touch. As the condition progresses, voluntary motion is lost and the fingers assume a flexed position. Passive extension of the fingers causes severe pain. The radial pulse is usually absent. The pain disappears after about forty-eight hours and contracture of the flexor group of muscles occurs with the subsequent development of a clawhand. Clinically, the presence of coldness, swelling of the fingers cyanosis and an absent radial pulse are most important early signs, however it must be noted that pallor rather than cyanosis is sometimes present and is equally as important. Pain in the forearm with the fingers assuming a flexed position and increased pain in the forearm on passive extension of the fingers is a serious sign, and if preventive treatment is to be attempted with any hope of success, earlier signs must be sought.

Treatment

The treatment of Volkmann's contracture is prophylaxis. The condition is easily missed in its earliest stages, and its recognition is essential if successful preventive measures are to be initiated. As a result of the injury the patient is expected to have some pain, and there is usually evidence of some swelling and tenderness of the fingers. Procrastination during this important stage is an easy matter and differentiation between normal discomfort and an impending Volkmann's contracture may be difficult. It should be remembered that following proper therapy of injuries such as fractures and dislocations the pain should rapidly subside. Cases with undue and prolonged pain should be watched with suspicion. All injuries about the elbow and forearm in children must be meticulously watched and preventive measures vigilantly initiated as soon as increasing swelling, cyanosis, coldness and pain appear. The time to treat Volkmann's contracture is in its very beginning. Once there has been degeneration and fibrosis with contracture of the muscle, there is no power that can restore the muscle to normal. From then on we must think in terms of improvement but never of cure.

Treatment in the Early Phase

As soon as the signs and symptoms of impending Volkmann's contracture appear one must first remove all dressings, bandages and splints and relieve the acute flexion of the elbow. One of the best methods of maintaining immobilization of the supracondylar fracture without acute flexion and without bandages or external splints is traction through a Kirschner wire in the olecranon. The use of repeated stellate ganglion blocks will frequently improve circulation. Should notable improvement appear with these measures resulting in the fingers becoming pink and warm and the return of a radial pulse, the case may be watched. If this happy state does not appear quickly within three or four hours surgical intervention is mandatory. Surgery is performed with the idea of relieving obstruction. The incision is made lengthwise over the flexor surface

of the forearm, jogging across the antecubital space to avoid crossing the flexion crease of the elbow The skin usually gaps as it is cut, and the deep fascia is found to be very tight as it is incised The brachial, radial and ulnar arteries are freed, and the muscle groups of the forearm are separated In making incisions through the deep fascia it must be remembered that the forearm consists of a series of deep fascial compartments The release of one compartment when another is involved is not the solution to the problem. It is important to *relieve the tension in each compartment. Closure is confined to the skin, and if* this cannot be accomplished without tension, a split thickness skin graft should be utilized.

Neurolysis of the ulnar and median nerves has been advocated and should be considered when primary involvement of the nerve appears to be present. At surgery, a few constricting bands or dense adhesions causing complete strangulation of the nerve may be discovered The nerve should be carefully freed by sharp dissection and saline neurolysis

Treatment in the Late Phase

The treatment of the established contracture is as varied as the theories of pathogenesis The reason for this is that the established contracture is not always the same Variable combinations and degrees of muscle involvement with or without nerve damage will present many types of contractures. Initial treatment during this stage should be conservative. In early cases gradual stretching by the method of Sir Robert Jones is excellent treatment. Using a rigid splint hinged at the wrist, the interphalangeal joints are first gradually extended then the metacarpophalangeal joints are extended and finally the wrist is brought into extension. The splint should be removed several times daily to institute active and passive exercises heat and massage to the hand. Rapid or forceful correction of the deformity produces overstretching laceration and hemorrhage inviting recurrence. There are many other methods of gaining gradual correction of the deformity by the use of bivalve casts semi rigid splints elastic traction splints glove traction splints and other pieces of apparatus The same general principle of gentle but persistent stretching of the deformity remains the same in every method.

In those cases in which there is sufficient viable muscle present to provide for practical function and muscle balance of the hand, passive stretching exercises and splinting may be the only treatment necessary In some instances, it may be found that certain structures are not stretching and are more deforming than valuable These structures should be sacrificed. Steindler states that the contracture resistance of muscle varies with the number of joints the muscle motivates. The short fibered uniarticular muscles of the wrist joint are very much more resistant in contracture than are the muscles of the fingers which have a wider contractile range because of the greater length of their muscle fibers The flexor carpi ulnaris offers the must resistance and lengthening or even sectioning of this tendon should be performed when necessary The same holds true for the flexor carpi radialis and the flexor pollicis longus.



Figure 294

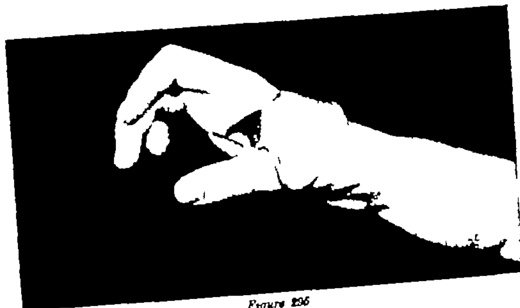


Figure 295

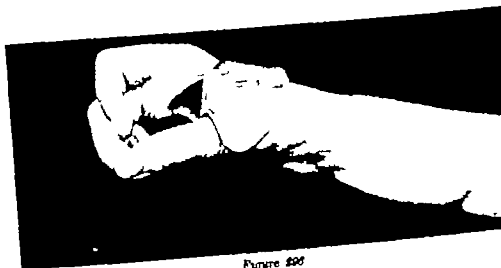


Figure 296



Figure 297



Figure 298

Figures 294, 295, 296, 297, 298. Same case as Figures 292, 293. Function of hand showing active (294) flexion (295) extension of fingers, (296) opposition, (297) writing and (298) grasping a glass, following multiple tendon transfers. (1) The extensor radialis longus and brevis were transplanted to the profundus and sublimis tendons after all of the flexor tendons have been released. (2) Brachioradialis transplanted into the flexor pollicis longus. (3) The extensor carpi ulnaris elongated by grafts from the long toe extensors and then transplanted through the intermetacarpal spaces to the lateral bands and (4) the extensor pollicis brevis was elongated by the palmaris longus tendon and rerouted around the ulna to replace opponents function. (This last procedure gave only a fair result but is not a good procedure due to the weakness of this muscle. Function is enhanced by use of an opponens splint.)



Figure 299 Volkmann's contracture of less severity This patient had some flexor profundus and sublimis function, but these were weak yet with contractures. Wrist flexors were strong but extensors poor



Figure 300 Same case as Figure 299 showing extension



Figure 301 Same case as Figures 299 300 Flexion following mobilization by physical therapy and transfer of the flexor carpi ulnaris to the base of the third metacarpal. This produced a dynamic tenodesis function whereby extending the wrist with the transfer allowed the tight finger flexors to grasp more or less passively

Reconstruction

A thorough knowledge of the mechanics and function of the hand plus the ability to assess the deformity with utmost care are necessary before attempting surgical reconstruction of the deformity caused by Volkmann's ischemic contracture Surgical procedures must be meticulously designed to salvage as much as possible from a serious deformity Excellent results are rarely obtained because of extensive damage to the functional mechanism of the hand These hands are cosmetically poor, however frequently some function can be obtained which will greatly decrease the handicap of the patient Occasionally, nothing can be done to improve function and the surgeon must be satisfied to accept this fact and be content with improving the hand if possible cosmetically The end results are innumerable, and each case presents an entirely different problem. This fact has resulted in many operative procedures being advocated for a single condition A procedure which is suitable for one type of problem may be quite unsatisfactory for another The general groups of procedures which are at present in common usage are (1) sliding of the flexor muscle origin distalward, (2) tendon lengthenings (3) shortening of static structures a) Shortening of the radius and ulna b) Carpectomy, (4) tendon transposition with or without carpal arthrodesis

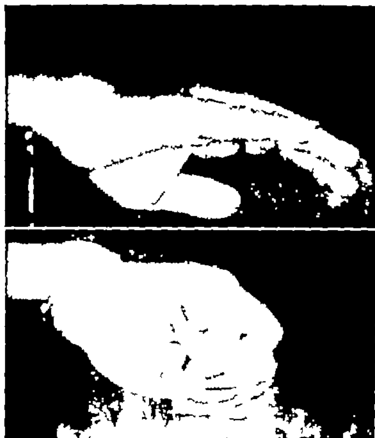
The first procedure produces a release of flexion contractures of the wrist, but detachment is possible for only a short distance because of the restriction of the median nerve branches Tendon lengthening of the wrist flexors is often helpful combined with stretching and other operative procedures Lengthening of the finger flexors is rarely of benefit Lengthening of these structures may allow the finger to be extended but it is usually found that by increasing the length the active contractile power of flexion has been diminished or lost in the functional range. Shortening of the long bones has decreased in popularity and it is now thought that if the actual shortening of the static parts will aid function, carpectomy is the procedure of choice In well selected cases this procedure will provide for good functional results and will frequently improve the cosmetic appearance of the hand. Tendon transplantations following a careful muscle evaluation will frequently aid in the functional improvement of the hand. This is especially true in the transplantation of extensors to the flexor muscles Capsulectomies of the metacarpophalangeal joints are often indicated, but not infrequently the contracture is so severe that the resection of the metacarpal heads or metacarpophalangeal arthroplasties will be necessary

The end results of the truly established cases of Volkmann's ischemic contracture are by no means excellent in anyone's hands. The recovery of some degree of function in these cases is often more appreciated by the patient than a brilliant correction of a mild deformity in another condition The rehabilitation of these hands presents a great challenge and offers great possibilities if a meticulous therapeutic plan is carried out.

References

BARDENHEUER E Deutsch, Ztschr f Chir 108: 44 1911

BROOKS B Experimental study of Volkmann's paralysis. Arch. Surg., 5: 188 1922





Figures 302 303 304 305 Function attained in a moderately severe Volkmanns' contracture after a carpectomy

Brook B Johnson C S and Kirtley J A, Jr Simultaneous vein ligation Surg Gynec & Obst 59: 496 1934
Bruce, J R Localized Volkmann contracture J Bone & Joint Surg 22: 738 1940
Burman M S and Sutro C J Brief communications experimental ischemic contracture Ann Surg 100: 550 1934
Cohen H H Adjustable volar-flexion splint for Volkmann's contracture J Bone & Joint Surg 24: 189 1942
D'Harcourt J and D'Harcourt M A contribution to the study of Volkmann's ischemic contracture Madrid Medicina 6: 237 1935
De Nuce Ischemic contracture Rev d'orthop., 20: 97 1909
Fleming C W Case of impending Volkmann's ischemic contracture treated by incision of deep fascia Lancet 2: 293 1931
Foisie, P S Volkmann's ischemic contracture New England J Med 226: 671 1942
Garber, J N Volkmann's contracture as complication of fractures of forearm and elbow J Bone & Joint Surg 21: 154 1939
Clex F W Vascular injuries complicating supracondylar fractures of humerus in children with case report Bull Jackson Mem Hosp 3: 29 1941
Griffiths D L Volkmann's ischemic contracture Lancet 2: 1339 1938
Griffiths D L Volkmann's ischemic contracture Brit J Surg 28: 239 1940
Harris C and Riordan D C Intrinsic contracture in the hand and its surgical treatment. J Bone & Joint Surg., 36A: 10 1954
Horwitz, T Significance of venous circulation about elbow in patho-mechanics of Volkmann's contracture Surg. Gynec & Obst 74: 871 1942
Jepson P N Ischemic contracture Ann Surg., 84: 785 1926
Jones, E B Volkmann's ischemia observations at open operation Brit M J 1: 1053 1940
Jones S G Volkmann's contracture Amer J Surg., 43: 325 1939
Jones R. Address on Volkmann's ischemic contracture with special reference to treatment. Brit M J 2: 639 1928.
Lagole, L Evolution of idea concerning Volkmann's syndrome Medicine 19: 893 1938
Massart R. Volkmann's disease ischemic contraction of the flexor muscles of the fingers Pathogenesis and treatment Presse med., 43: 1695 1935
Meyerding H W Fracture of elbow and Volkmann's ischemic contracture Minn. Med., 22: 100–104 1939
Meyerding, H W Volkmann ischemic contracture Jour Amer Med Assoc., 94: 394–400 8 1930
Meyerding, H W Volkmann's ischemic contracture Surg. Clin. N Amer., 10: 49–60 1930
Meyerding H. W and Krusen F H Treatment of Volkmann's ischemic contracture 110: 417–426 1939
Middleton D S Discussion of Volkmann's ischemic contracture Lancet, 2: 299 Aug 1928
Murphy J B Myositis. J A M A 63: 1249 Oct. 1914
Plewes L W Occlusion of brachial artery and Volkmann's ischemic contracture Brit. Med. Jour 1: 1054–1055 June 29 1940
Powers C A The ischemic paralysis and contracture of Volkmann. J A M A 48: 759 Mar 1907
Pruitt M E Abortive treatment of Volkmann's ischemia. Jour Kansas Med Soc., 35: 448–451 Dec. 1934
Rowlands Lancet 2: 1168 1905
Steindler A. Ischemic contracture Surg. G & O No 2A, 62: 358–364 Feb 1936
Volkman Pitha, and Billroth Handbuch der Chirurgie, 2: 846 1882
White, J W., and Stubbins S G Carpectomy for intractable flexion deformities of the wrist. J Bone & Joint Surg 26: 131–138 Jan. 1944.

WRINGER ARM

Wringer arm is a descriptive term used when the upper extremity is caught between two rollers. These injuries occur most frequently in childhood and are usually produced by power driven washing machines. Hardin and Robinson reported 35 cases of which 30 were children under the age of 6 years.

Pathogenesis and Pathology

The compressive force and friction produced by the rollers cause damage primarily to the skin and soft tissues of the hand, forearm and elbow region. The severity of damage depends upon the length of time spent between the rollers, the rapidity of revolution width of approximation and tension of the rollers. The prognosis is much better in the younger age groups because of the elasticity and flexibility of the bones joints and ligaments

In these injuries, the hand usually enters the roller first with the fingers in the extended position. There is usually enough give in the tension of the rollers to allow the hand and the distal portion of the forearm to pass through without too much damage As the forearm proceeds through the roller, tension usually builds up because of the increasing width of the upper arm and elbow, and this area usually bears the brunt of the avulsing action of the rollers.

Bones are rarely damaged, but muscles, tendons and nerves may be crushed, torn or avulsed without a break in the skin. The most common damage is that of separation of the skin from its subcutaneous attachments by the mechanical action of the moving rollers. A collection of blood and serum beneath such a full thickness skin area delays the reestablishment of good blood supply If this condition goes unrecognized and untreated the overlying skin remains separated and sloughs. Venous thrombosis and lymphatic obstruction occur frequently The arteries in the affected zone are less easily injured than the veins and continue to circulate blood into the damaged area causing engorgement and subsequently necrosis Occasionally flaps of skin are completely separated exposing contused or macerated muscle. At times tendon ends are completely stripped from their muscle attachments.

Clinical Examination

Immediate evaluation should include the determination of nerve and tendon function distal to the site of the injury and roentgenographic examination for possible bone or joint damage. In those cases seen immediately after injury, the swelling and ecchymosis may be mild to moderate in amount, misleading the physician inexperienced in treating these conditions. Occasionally, the damage may be limited to the skin with only abrasion, contusion and edema being present. In many instances however extravasation of blood and serum separate the skin as a flap from the forearm or arm and severe sloughs occur The extent of damage cannot always be determined early and the prognosis should always be guarded.



Figure 300 Wringer injury Besides deep slough shown here the child has paralysis of the radial, ulnar and median nerves

Treatment

Regardless of the initial appearance of the injury the patient should be hospitalized and observed for at least 48 hours Conservative treatment should be employed in which viable skin covers the damaged area. Pressure dressings and elevation of the extremity will minimize the edema of the damaged limb The area should be cleansed with soap and water, preferably Phisoderm with hexachlorophene and dressed with sterile dressings. The use of pressure dressings applied as one dresses a new skin graft will often prevent separation of the skin from the underlying tissues. This dressing should be changed in 48 hours and if a hematoma has lifted the skin aspiration should be performed, and the pressure dressing reapplied In late cases with severe extravasation the collection of blood should be aspirated or released by multiple small incisions and then dressed with pressure dressings with subsequent elevation and observation.

Definitive reconstructive surgery should be deferred until all inflammatory induration has subsided and the degree of soft tissue destruction can be ascertained Good surface coverage should be completed in all cases before tendon and bone reconstruction is attempted If the injury has produced a flap, resuture will usually be unsuccessful. Farmer has successfully excised the fat and subcutaneous tissues converting the flap into a full thickness graft. The depth of injury will limit this type of treatment because of the necessity of a suitable underlying bed. A split thickness graft should be used when there is permanent loss of

skin but this will usually result in shrinkage and secondary coverage should be carried out. If bone tendon joint or nerve is exposed an immediate pedicle graft is imperative Rotation or sliding flaps are usually unsuccessful because of tissue damage and circulatory impairment to the immediate surrounding tissues

The thoracico-abdominal area is a convenient source for covering these severely damaged areas as described by Hardin and Robinson These flaps are developed conventionally with a superior base and should be one-third larger than the defect to allow for shrinkage The length should not exceed the base by more than two times. The flap is carried to the external oblique fascia and is transferred without any removal of fat. The abdominal defect may be closed by undermining or if too large, coverage may be completed with a split thickness graft. The graft is dressed with moderate pressure and immobilized to relieve any motion or tension on the graft base. Detachment and revision is carried out in approximately three weeks. If tendon repairs or grafts are necessary, all joints should be thoroughly mobilized passively prior to surgery The skin should be well healed with subsidence of induration before definitive work is attempted in order to prevent summation of scarring.

References

DUPERTUIS, S M An evaluation of skin grafts for hand coverage J Bone & Joint Surg., 34-A: 811–819 Oct. 1952.

FARMER, A. W., Treatment of avulsed skin flaps, Annals of Surg., 110: 951–960 1939

HARDIN C A. AND ROBINSON D W., Coverage problems in treatment of wringer injuries, J Bone & Joint Surg., 36-A: 292–298, Apr 1954.

MACCOLLUM, D W., Wringer arm a report of twenty-six cases, New England J of Med 218: 549–554 1938

MACCOLLUM, D W BERNHARD W F AND BANNER, R L *The treatment of wringer arm injuries*, New England J of Med 247: 750–751, 1952

POSCH J L. AND WELLER, C N Mangle and severe wringer injuries of the hand in children. J Bone & Joint Surg., 36-A: 57–63 Jan. 1954.

LACERATIONS OF THE HAND AND FINGERS

Damaged tendons may occur as a result of lacerations, burns, crushing injuries, friction wounds or combinations such as seen in wringer or mangle injuries discussed elsewhere in this chaptre. Most lacerations occur in the fingers due to grasping sharp cutting surfaces. The treatment of these wounds varies in no way from the treatment in adults Boyes has grafted tendons in two-year-old children with excellent results The tendons grow in length and continue to function well with growth. Because of the smallness of the parts the difficulty with immobilization and poor cooperation it is thought best to wait until the child is at least two years of age before performing tendon grafts. Joints should be kept mobile by passive exercise until surgery can be performed. No tendon surgery should be done until good skin coverage has been attained and all scars have been released. The bony parts should be intact, and all joints of the involved digits should be passively mobilized before tendon surgery

If the profundus tendon has been cut distal to the sublimis attachment, the



Figure 307 Attempted flexion after severance of the flexor sublimis and profundus in an eleven year-old boy

Figures 308 309 Same case as Figure 307 Range of flexion after removal of the sublimis and profundus tendons and tendon graft using palmaris longus to the profundus in the palm extended to the base of the distal phalanx.

distal end of the cut profundus can be advanced to a bony attachment at base of the distal phalanx. The distal piece of the cut tendon is then remo The suture is carried out by using a stainless steel pullout wire suture atta into the bone at the base of the distal phalanx. The tendon will soon accom date to its new length and a satisfactory functional result is obtained. If or both tendons are cut in the area on the volar surface between the sublimi *sertion and the distal palmar crease both tendons should be removed. A g* should be placed from the profundus tendon in the palm out through the fi to a bony attachment into the proximal end of the distal phalanx. This is c according to the method of Bunnell using #34 stainless steel wire sutures pulleys have been destroyed, they are replaced, using tendons wrapped al the phalanges or metacarpal necks and passing subcutaneously on the do surface outside the extensor apparatus. The suture line in the pulley shoul fixed in an area not in contact with the tendon graft. The hands are immobil routinely in plaster splints to relieve tension on the graft for a period of t weeks Following removal of the cast, active exercises and physical therapy started. If a tendon seems to be tied down a tendolysis may be necessary, one should not be hurried in performing such a procedure in a child, inasmuc adhesions if they occur will often stretch out after several months of active passive exercises. In the majority of instances, the tendon begins to function within the first three or four weeks after removal of the casts or splints.

Lacerations occurring in the flexor aspects of the fingers are best treated debridement, cleansing and skin suture, leaving the graft to be done later as elective procedure. In those lacerations involving the extensor aspects of hand and fingers, the extensor tendons should be sutured primarily

Children are not infrequently seen with deformities of the hand and fin due to unrecognized lacerations of the peripheral nerves higher in the arm forearm. Many of these lacerations have been treated as simple, small cuts w out due recognition of the nerve damage Fixed contractures that have occur should be corrected if the injuries are seen late, and the nerves should then explored just as in the adult patient, suturing the ends carefully with #6-0 on an atraumatic needle. If this procedure is carefully performed, a great nu ber of these children will regain full function. Satisfactory results may be pected as late as two years after the injury for return of motor function and e later for the return of sensation. Digital nerves damaged in the hand and fing should be sutured at the time of debridement, *using a careful, meticulous te* nique with #6-0 silk on an atraumatic needle.

If motor function does not return there are various reconstructive transpla which can be performed with good results. In the case of a radial nerve loss, go results are usually obtained by transplanting the wrist flexors into the finger a thumb extensors. The most common procedure is the use of the flexor ulna into the common extensors, the palmaris longus into the abductor and exten brevis of the thumb and the flexor carpi radialis into the extensor pollicis long There are other methods of utilizing these same muscles just as effectively l each procedure depends upon individual variations.

Extensor tendon
Bone
Incision
Neurovascular bundle
Flexor tendon

Figure 310 Incisions useful in the hand

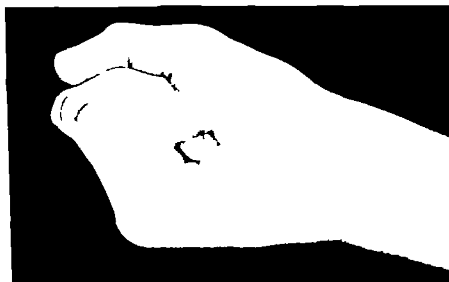


Figure 311 Severed profundus tendon in the right little finger between the proximal and middle crease of the finger One slip of the sublimis had also been severed, and there was deep scarring and destruction of this pulley

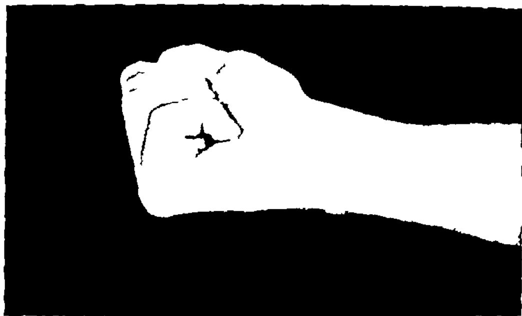


Figure 312 Same case as Figure 311 Show flexion and extension following tendon graft. The sublimis and profundus tendons were removed The palmaris longus was grafted from the profundus in the palm through the finger to the base of the distal phalanx. The proximal pulley was reconstructed with a part of the removed sublimis tendon.

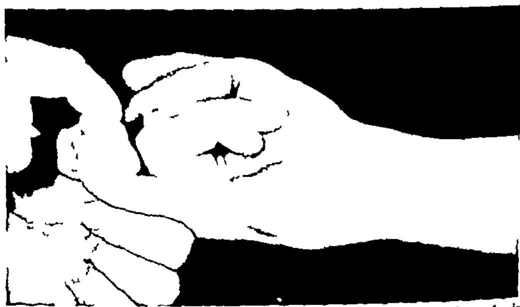
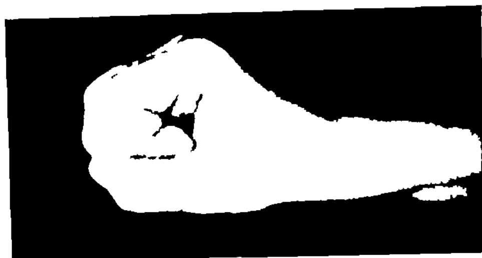
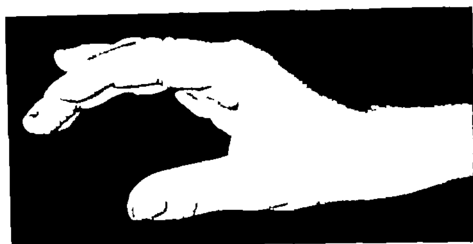


Figure 313 Same case as Figures 311 312 Showing the definite function of the profundus and its strength after the profundus graft







Figures 316–318 Same case as Figure 314 Function of hand following transplantation of flexor carpi radialis to extensor pollicis brevis and abductor pollicis longus palmaris longus to extensor pollicis longus and flexor carpi ulnaris to common finger extensors.

In ulnar and median paralyses, the main loss is that of intrinsics. The consequent loss of finger extension and thumb opposition is quite disabling. The different varieties of transplants that may be used to restore these functions will not be enumerated. Deformities can be overcome completely or partially by appropriate tendon transfers and arthrodeses designed to restore muscle balance and bring the thumb and fingers to functional positions. Often there is a dearth of transferable tendons so that only the most important intrinsic function can be restored. In children it is best to avoid arthrodeses if possible. Irwin's dynamic tenodesis may give adequate function by utilizing a few tendons and retaining joint motions. Occasionally injury will result in the necessity of sacrificing digits or a portion of the hand. Such a loss is always disabling, however, the axiom of preserving or restoring the thumb at all costs should always be kept in mind by the surgeon.

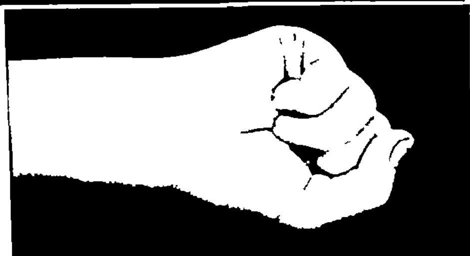
Sometimes parts of fingers or entire fingers must be sacrificed. The loss of a finger is disabling but not nearly as disabling as the loss of a thumb. In instances of loss of the terminal part of the thumb by simply performing the procedure of deepening of the web space by a Z-plasty, the function may be greatly improved. Sometimes when both phalanges are lost while the first metacarpal is retained along with its musculature, certain procedures may be performed to increase the length and usefulness of the thumb. Philip Lewin has described an advancement procedure using a bone graft into the metacarpal and a skin graft on a nonoppos-



Figure 318 Radial nerve palsy due to trauma







Figures 321 322 323 Same case as Figure 320 Result two years after ulnar nerve suture Patient has complete recovery of intrinsic function and sensation.

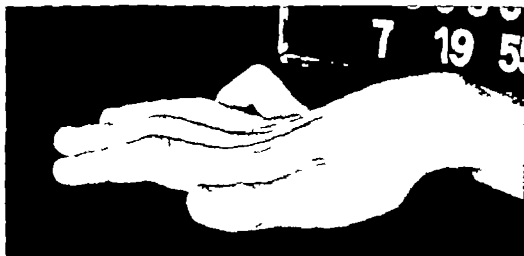
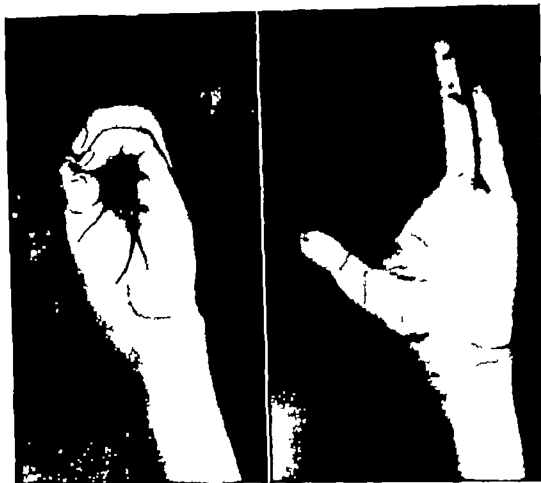


Figure 319 Ulnar nerve palsy due to laceration over the flexor surface of the ulnar side of the elbow This is only five months old and the wound is well healed No finger contractures are present. The nerve should be explored and sutured at this time.



Figure 320 Attempt at extension of fingers after laceration at the ulnar surface of the wrist which severed the ulnar nerve at that level with loss of the intrinsic function due to the ulnar nerve.



Figures 326 327 After bone grafting the second metacarpal into the first metacarpal through a pedicle graft then release of the pedicle and closure Good functional and cosmetic improvement.

Tube pedicles may be constructed and a bone graft equal to the length of the previous thumb moved into the pedicle and grafted into the first metacarpal The motor function of these reconstructed parts is usually excellent, but the sensation is poor, making its usefulness inadequate This inadequacy may be solved now, however, by using the skin from a less valuable finger such as the little or ring finger The skin is taken from the flexor surface at the tip and transferred with its neurovascular bundle according to the method of Littler to the opposing end of the thumb This addition should make for much better functional results, but great care is necessary in performing this type of surgery

References

ABBOTT L. C SAUNDERS J B AND BOST F C Arthrodesis of the wrist with the use of grafts of cancellous bone J Bone & Joint Surg., 24: 883 1942.

BOYES J H. Repair of motor branch of the ulnar nerve in the palm J Bone & Joint Surg., 37A: 920 1955.

BOYES J H Flexor tendon grafts in fingers and thumb J Bone & Joint Surg., 32-A: 489 1950

BOYD H B AND STONE, M M Resection of the distal end of the ulna. J Bone & Joint Surg., 26: 313 1944

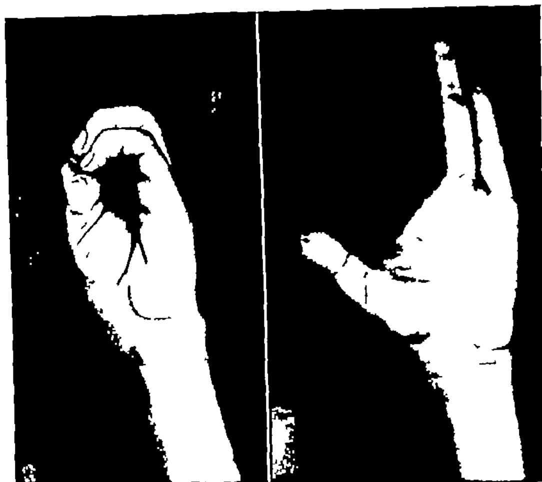
ing surface, lifting the intact skin up over the bone graft and inlaying the skin below the surface This procedure may give some added length with sensation on the opposing surface and serve greatly for the previous loss. Littler has gone even further using almost the same principle described by Lewin except when the thumb is retained, Littler uses a procedure to improve the total function of the hand when all of the fingers including the metacarpals have been lost elevating the skin flap and inserting a bone graft from the ilium underneath the skin flap, using the normal skin from the hand with its sensation as the part to oppose against the thumb and then filling the part that is not to be opposed with a skin flap from the abdomen. This will give an effective post for the intact thumb to work against, this post has good sensation on the part that the thumb will oppose against.



Figure 324 Loss of hand except for the thumb due to trauma. Would benefit greatly by procedure described by Littler to give a post for the thumb to work against.



Figure 325 Loss of phalanges of the thumb index and middle fingers due to trauma.



Figures 326 327 After bone grafting the second metacarpal into the first metacarpal through a pedicle graft then release of the pedicle and closure Good functional and cosmetic improvement

Tube pedicles may be constructed and a bone graft equal to the length of the previous thumb moved into the pedicle and grafted into the first metacarpal The motor function of these reconstructed parts is usually excellent, but the sensation is poor making its usefulness inadequate This inadequacy may be solved now, however, by using the skin from a less valuable finger such as the little or ring finger The skin is taken from the flexor surface at the tip and transferred with its neurovascular bundle according to the method of Littler to the opposing end of the thumb This addition should make for much better functional results, but great care is necessary in performing this type of surgery

References

Abbott L. C Saunders J B and Bost F C Arthrodesis of the wrist with the use of grafts of cancellous bone J Bone & Joint Surg 24: 883 1942.

Boyes J H Repair of motor branch of the ulnar nerve in the palm. J Bone & Joint Surg., 37A: 920 1955

Boyes J H Flexor tendon grafts in fingers and thumb J Bone & Joint Surg., 32-A: 489 1950

Boyd H B and Stone, M M Resection of the distal end of the ulna. J Bone & Joint Surg., 26: 313 1944

BRUNER, J M Problems of postoperative position and motion in surgery of the hand. J Bone & Joint Surg 35A: 355 1953

BUNNELL, S Active splinting of the hand. J Bone & Joint Surg., 28: 732 1946

BUNNELL, S Digit transfer by neurovascular pedicle J Bone & Joint Surg 34: 772, 1952.

BUNNELL, S. Physiological reconstruction of the thumb after total loss. Surg. Gynec. & Obst., 52: 245 1931.

BUNNELL, S. Primary and secondary repair of flexor tendons of hand. J Am Soc Plastic & Recon. Surg 12: 65 1943

BUNNELL, S Repair of tendons in the fingers and description of two new instruments. Surg. Gynec & Obst., 26: 103 1918

BUNNELL, S Surgery of the intrinsic muscles of the hand other than those producing opposition of the thumb J Bone & Joint Surg., 24: 1 1942

BUNNELL, S Suturing tendons. Amer Acad. Orthoped. Surg., Lect.1-5 1943

BUNNELL, S Treatment of tendons of the hand. J Bone & Joint Surg., 23: 240 1941

BUNNELL, S Surgery of the hand J B Lippincott Co Philadelphia, 1944

DIAL, D Reconstruction of the thumb after traumatic amputation. J Bone & Joint Surg., 21: 98 1939

DUNLOP J The use of the index finger for the thumb some interesting points in hand surgery J Bone & Joint Surg 5: 99 1923

GOLDNER J L. Deformities of hand incidental to pathological changes of the extensor and intrinsic muscle mechanisms. J Bone & Joint Surg 35A: 115 1953.

GRAHAM W C Flexor tendon grafts to the fingers and thumb. J Bone & Joint Surg., 29: 553 1947

JOYCE, J L. Results of new operation for substitution of the thumb Brit. J Surg., 16: 362 1929

KOCH S L. AND KANAVEL, A B Purposeful splinting following injuries of the hand. Surg. Gynec. & Obst., 68: 1 1939

LEWIN MICHAEL Partial reconstruction of the thumb in a one stage operation. J Bone & Joint Surg 35A: 573 1953.

LITTLER J W Architectural principles of reconstructive hand surgery Surg. Clinics of North America, 31: 463 1951

LITTLER, J W Free tendon grafts in secondary flexor tendon repair Am. J Surg 74: 315, 1947

LITTLER, J W Tendon transfers and arthrodeses in combined median and ulnar nerve paralysis. J Bone & Joint Surg., 31A: 225 1949

LITTLER, J W The neurovascular pedicle method of digital transposition for reconstruction of the thumb Plast & Reconstruc. Surg 12: 303 1953

LITTLER J W Transfer of free skin graft with neurovascular pedicle (Presented at 1956 meeting Am. Society for Surgery of Hand)

RIORDAN D C Tendon transplantation in median nerve and ulnar nerve paralysis. J Bone & Joint Surg 35A: 312 1953

SILER, V E. Primary tenorrhaphy of the flexor tendons in the hand. J Bone & Joint Surg., 32A: 218 1950

BURNS

Burns involving the hands are unfortunately common injuries in children. *Very frequently other parts of the body are involved greatly complicating the* overall treatment program. The burn usually involves only the skin, sparing the tendon bones and joints however if treated improperly in the earlier stages involvement of these structures may occur secondarily with severe crippling disability of the hand. The types of deformity produced may take almost any form and are often quite grotesque The most common deformity is that of

flexion of the wrist, hyperextension of the metacarpophalangeal joints and flexion of the interphalangeal joints, or flexion of the proximal interphalangeal joints with extension of the distal joints These types of deformity tend to be more frequent since pain and muscle spasm cause overpull of the wrist flexors with stretch on the common extensors This causes a hyperextension of the proximal finger joints which often results in lateral slipping of the lateral bands to produce severe finger deformities. Sometimes these burns may be severe enough to cause a loss of major deep structures primarily such as ligaments and tendons Occasionally, fingers or parts of fingers or practically the entire hand will be lost If the wounds are allowed to become septic or there is an accumulation of necrosis with poor debridement joints and tendons will become fixed rapidly and often permanently The skin of children and infants is relatively thin and deeper involvement may occur initially than would have occurred in an adult with a similar type burn Fortunately, the joints of a child do not stiffen quite as rapidly as those of the adult Children do not tolerate burns in general very well during the initial phases

Treatment

The systemic treatment of burns does not differ in children from the therapy indicated in adults The general care of toxemia shock and sepsis, all of which are of great importance will not be discussed here. An attempt will be made



Figure 323 Burn healed by granulating in without any skin coverage showing severe cicatrix contracture

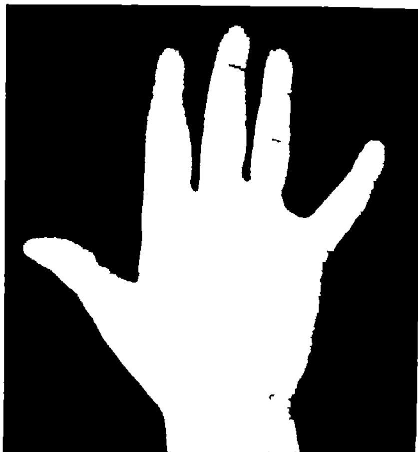


Figure 329 Same case as Figure 328 after excision of scar and replacement by full thickness skin graft from the abdomen.

to outline and cover the more important aspects of the local care of the hand. It is essential that all efforts should be made to allay pain, prevent sepsis, debride early and cover as soon as possible with skin grafts Splinting in the position of function is extremely important to prevent deformity

Early Therapy

Methods of early treatment are controversial, but the majority of authorities are of the opinion that open surgical drainage is the treatment of choice Hand burns seen in the first twelve hours or even those seen after sepsis has occurred should be treated in the following manner with the general idea of gaining skin coverage as soon as possible. This can be obtained by early debridement, daily cleansing and wet dressings. As soon as the general condition permits, the entire hand should be cleansed gently and thoroughly with large gauze sponges using hexachlorophine and phisoderm with water The blisters should be opened and necrotized and charred tissue removed. This procedure should be exacting and meticulous. Several layers of fine mesh gauze should be wrapped around the part, dressing the fingers separately and being careful to place each in the position of function with the wrist dorsiflexed fingers in semi flexion and the thumb slightly flexed and in opposition. Sterile mechanic's waste may then be

applied to hold these positions and the entire bulky dressing kept wet with normal saline solution. Metal or plastic splints may be incorporated to hold position when necessary, but usually the bulky mechanic's waste dressing will suffice to hold the position satisfactorily The dressings are changed under water, and the patient is encouraged to move his fingers actively as much as possible Necrotic tissue is trimmed away as it is encountered. The majority of hand cases can be prepared in this manner for split thickness coverage within two to four weeks In most instances, this original covering of split thickness skin will serve permanently on the dorsal aspect of the hand and fingers. On the palmer aspect of the hand and fingers, the split graft is used only for temporary coverage It is later replaced with full thickness skin grafts.

Late Treatment

Unfortunately, many burned hands are seen late with severe scarring and contractures No burn wound should be allowed to "granulate and heal in" Every burn should be covered as soon as possible.

When scarring of the dorsal surface of the fingers occurs the scar can be completely dissected free the fingers flexed and immobilized on an aluminum splint. The area can be covered with a split thickness skin graft. These aluminum splints can be prepared and cut to shapes as necessary prior to surgery and then sterilized and molded into the positions desired in the operating room. Pressure dressings are applied over the graft and the entire part dressed with bulky pressure dressings incorporating the splint The dressings are changed in about ten days and immobilization is continued for approximately one month then mobilization is started using physical therapy in the form of active exercises and whirlpool The splint is used on a part time basis until the end of the sixth week. Scars involving the palm should be removed and the hand splinted with the fingers in extension after a full thickness graft has been applied These grafts are obtained from an area on the abdomen containing as little hair as possible.

The graft is outlined by a pattern and the dissection is carried out with the graft under tension following a plane in the fine white fibers which attach the skin to the subcutaneous tissue No fat is removed with the graft The graft is cut slightly larger than the wound to allow for shrinkage It is sutured accurately into the bed which should be perfectly dry in order to prevent hematoma formation. A central holding stitch or two is often used to prevent slippage

The donor site can usually be closed primarily by excising the subcutaneous fat undermining and closing the defect. If the defect is too large a split thickness graft may be necessary to cover without tension. In most instances it is necessary to immobilize the elbow in flexion attaching this part of the cast to the splint to keep it from slipping. Longitudinal scars involving the flexor surfaces of the finger and web spaces are not uncommon in burns of the hand. They may be released by Z-plasty or multiple Z-plasties or by cutting the longitudinal scar transversely allowing the finger to extend and then inserting a diamond shaped full thickness graft. The release of tension by these methods results in the gradual disappearance of the scar and contracture.

Scar
Incision
B.
C
Free Graft
A.
D

Figure 330 Diagram of procedure for release of scar



Figures 331, 332 Longitudinal cicatrization after burn. This may be released by a Z-plastic procedure with an excellent result.

Dry fields are essential Immobilization as described above is mandatory When final coverage for hands and fingers has been secured and mobilization has been attained fixed deformities may remain due to tendon, bone or joint damage If the middle extensor slip of the extensor apparatus scars down, the lateral bands slip laterally, causing flexion of the middle joint and extension of the distal joints, the middle slip will be freed and the lateral bands sectioned. This procedure allows the distal phalanx to flex into the mallet finger position but allows for better function in the proximal and middle joints In severe cases the joint surfaces may be extensively damaged and the tendons and ligaments scarred beyond repair Arthrodesis in a functional position may be the treatment of choice Occasionally, an amputation may be most desirable This decision will depend upon the individual case If flexor tendons have been lost, they may be replaced by grafting as in other traumatic flexor tendon injuries. (See Lacerations) Frequently the results are rather discouraging because of the other involvement such as joint tightness and trauma to the capsule by the burn itself Tendon grafts should not be undertaken until good skin coverage has been secured in the area of transplantation It is also imperative that a maximum functional passive range of motion is attained in the involved joint before the tendon work is started. In many instances only partial restoration is possible however, because of the serious disability which occurs as a result of severe burns to the hand reconstruction should always be attempted in order to gain at least some function and possible cosmetic improvement

References

BARSKY A J., Surgical repair of unusual dermatological conditions, Surg. Clinics of N A., 19: 459–466 Apr 1939

BLAIR, V P., AND BROWN J B Use and uses of large split grafts of intermediate thickness, S G & O 49: 82–97 July 1929

BROWN J B Surface defects of the hand Am. Jour of Surg., 46: 690–699 Dec. 1939



Figure 333 Severe scarring following a burn.



Figure 334 Same case as Figure 333 after freeing of scar and application of a full thickness graft, releasing the thumb individually and the fingers *en masse* from the palm

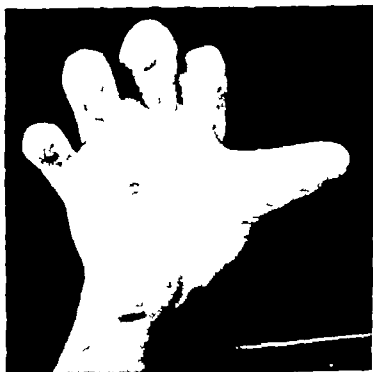


Figure 335 Same case as Figures 333 334 after releasing the webs between fingers as one would perform in syndactylism





Figure 336 Flexor cicatrix due to old burn *Figure 337* Same case as Figure 336 after release of scar and coverage by diamond inlay of free full thickness skin from the abdomen.



Figure 338 Showing result of poor early handling of a burn. No splinting. Healing has occurred by granulating in producing heavy cicatrization with inexcusible deformity due to contracture



Figure 839 Soon after burn due to friction. This child grasped a belt of a rapidly rotating motor

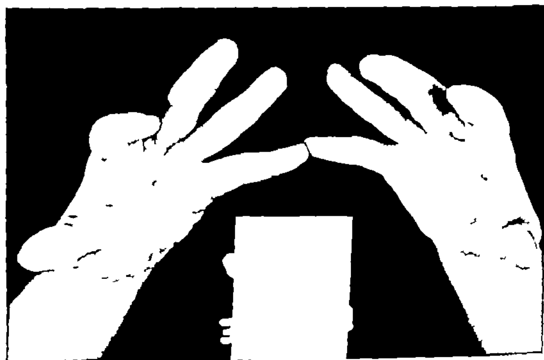


Figure 840 Free full thickness grafts used in the palm to replace cicatrices



Figure 3.1 Severe burn of hand and wrists mainly on the dorsal surfaces with fixed hyperextension contractures of the metacarpophalangeal joints and flexion contracture of the proximal interphalangeal joints and hyperextension of most of the distal interphalangeal joints.



Figure 3.2 Same case as Figure 3.1 Active splinting used to help relieve some of the contractures.

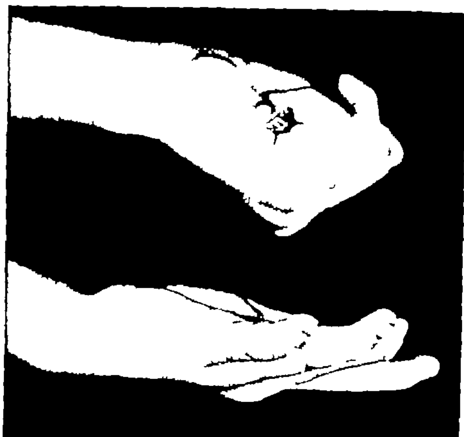


Figures 343 344 Same case as Figures 341 342 after removal of the dorsal scar of the wrist, hand and fingers to the proximal interphalangeal joints and coverage with split thick ness grafts. Release of the distal extensors of the lateral bands may improve the fingers by releasing the extension contracture of the distal joint and the flexion of the proximal inter phalangeal joint which have occurred through buttonholing of the lateral bands.

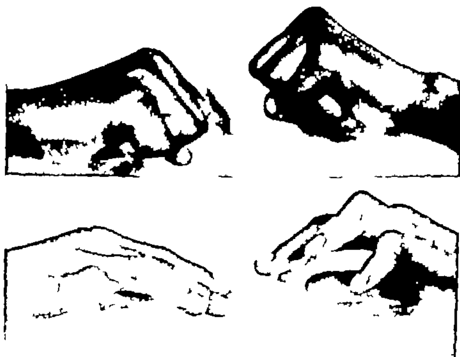
Figure 345 Flexion contractures in volar skin of all fingers. *Figure 346* Same case as Figure 345 after a diamond free full thickness skin graft.



Figure 347 Flexion contracture of the ring and middle finger due to a burn The skin only was involved on the ring finger The tendons and the proximal interphalangeal joint were scarred in the middle finger Both fingers were released and covered with a free full thickness skin graft Later the middle finger was covered by a pedicle flap from the abdomen and a free tendon graft was inserted using the palmaris longus tendon from the profundus tendon in the palm to bony attachment at the base of the distal phalanx.



Figures 348 349 Same case as Figure 347 Result after treatment four years later (In an adult, amputation of the middle finger would have been the procedure of choice.)



Figures 350 351 Result after release of a severe cicatrix in the palm due to burns. There is still flexion tightness of the skin in the fingers which will be released and fill by free full thickness graft just as in the palm.



Figure 352 (left) Severe burn with contracture of the middle finger and loss of the ring and little finger in a girl of seven years. *Figure 353 (right)* Same case as Figure 352 after release of the scar and removal of the fourth and fifth rays using some of the skin from the fillet to cover the scarred areas.

BROWN J B AND McDOWELL, F Persistence of function of thin grafts through long periods of growth S G & O **72**: 848-853 May 1941
BROWN J B AND McDOWELL, F Skin grafting of burns. J B Lippincott Co., 1943
BUNNELL, S Contractures of the hand from infection and injuries J Bone & Joint Surg., **14**: 27-46 Jan. 1932
BUNNELL, S AND HOWARD L. D Additional elastic hand splints, J Bone & Joint Surg **32-A**: 226-228 Jan. 1950
COHEN H. H An adjustable volar flexion splint J Bone & Joint Surg **24**: 189-192 Jan. 1942
CONWAY H., Whole Thickness grafts in correction of contractures due to burn scar Annals of Surg., **109**: 286-290 Feb 1939
DAVIS, J S Use of small deep grafts in repair of surface defects, Am. J of Surg., **47**: 280-298, Feb 1940
DUPERTUIS, S M An evaluation of skin grafts for hand coverage J Bone & Joint Surg., **34-A**: 811-819 Oct. 1952.
GILLIES H., Experiences with tubed pedicle flaps, S G & O **60**: 291-303 Feb 1935
GOLDNER, J L., Deformities of the hand incidental to pathological changes of the extensor and intrinsic muscle mechanism J Bone & Joint Surg., **35-A**: 115-131 Jan. 1953
HARRIS C AND RIORDAN D C Intrinsic contracture of the hand and its surgical treatment, J Bone & Joint Surg **36-A**: 10-20 Jan 1954.
HOVENS F Z Preoperative and postoperative care in skin grafting, Surg Clinics of N A **20**: 1,087-1,092 Aug 1940
HOWARD L. D JR., Contracture of the thumb web J Bone & Joint Surg **32-A**: 267-273 Apr 1950
JONES R., JR., Management of old contractures of upper extremity resulting from third degree burns, Sou Med. Jour **4**: 789-797 Aug 1911

KOCH S L., Complicated contractures of the hand Annals of Surg 98: 546-580 Oct. 1933
KOCH S L. AND KANAVEL, A. B Purposeful splinting following injuries of the hand, S. G & O 68: 1-16 1939
MASON M L., Plastic surgery of hands, Surg Clinics of N A 19: 227-248, Feb 1939
PADGETT E. C., Free full thickness skin graft in correction of soft tissue deformities J A. M A., 98: 18-23 Jan. 1932
PADGETT E C. Skin grafting, Charles C Thomas, Springfield, Ill 1942
PADGETT E. C., *Skin grafting in severe burns, Am Jour of Surg., 43: 626-636 Feb 1939*
PADGETT E. C., Skin grafting and three-quarter thickness skin graft for prevention and correction of cicatricial formation, Annals of Surg., 113: 1,034-1,049 June 1941
SHEEHAN J E., Use of free full thickness skin grafts, J A M A., 112: 27-29 Jan. 7 1939.
TANZER R. C Prevention of postoperative hematoma in surgery of the hand, use of compression suture J Bone & Joint Surg 34-A: 797-804 Oct. 1952



Figures 354 355 Severe deformity of the hand due to cicatrix of the skin involving mainly the dorsal aspect.



Figures 356 357 Condition after release of scar and free full thickness grafts from the abdomen



Figure 358 Severe burn with loss of all fingers The first metacarpal remained with its intrinsic muscle attachment intact.

BRACHIAL PALSY—BIRTH TYPE

A paralysis due to injury of the roots of the brachial plexus at birth became a more common clinical entity as fetal and maternal mortality associated with difficult deliveries was reduced. Still further refinement of obstetrical technique has reduced the incidence in recent times

Smellie mentioned this complication of delivery in 1764 The mechanism of the paralysis with nerve root injury was described by Duchenne in 1872 and Erb in 1874. A predominant upper arm lesion bears their name Klumpke described the lower arm type in 1885

Etiology

Fixation of the shoulder or head followed by traction and lateral flexion of the neck by a lever arm formed by either trunk arm or head has been felt to be the mechanism of injury



Figures 359 360 Condition after a Z-plastic procedure with deepening of the web between the first and second metacarpal. The child now has a much improved function.

Pathology

In a review by Wickstrom *et al* in fifty four patients, thirty six had their involvement centering about the fifth and sixth cervical roots In this group of principally upper arm involvement, minimal damage was most common This would be represented at the root by edema and mild intraneural hemorrhage, some tearing of fibres, but with the root still intact In this group recovery might occur in a few days, but more commonly complete return is evidenced in three to six months

There were fourteen cases in whom all roots were involved and four in whom paralysis was evidenced as involving primarily the eighth cervical and first thoracic roots

In these there is disruption of the nerve fibres at multiple points This includes avulsion of the roots from the spinal cord Complete laceration of the roots, multiple neuromata and gross lesions of the cord have been observed on exploration.

There were three cases of Horner s syndrome and four with sensory as well as motor loss in Wichstrom's series

Clinical Picture

Early

The failure of the involved arm to achieve the same degree of activity as the uninvolved limb is striking in early post natal life The stricken limb tends to remain lifeless at the side with elbow extended. Pronation of the forearm and wrist strap may be added placing the limb in the attitude of receiving a "hidden tip

The lifeless attitude is true of those cases having minimal as well as those having severe damage in early post natal life.

There may be swelling in the supraclavicular fossa due to hemorrhage and inflammation Occasionally a fractured clavicle or humerus is seen in concert with the paralysis. Signs consistent with Horner s syndrome may be found if the first thoracic root has been injured

Late

The signs of this injury in the older child are those of residual deformity The most marked imbalance of musculature is that of strong internal rotators at the shoulder as compared to paralyzed external rotators

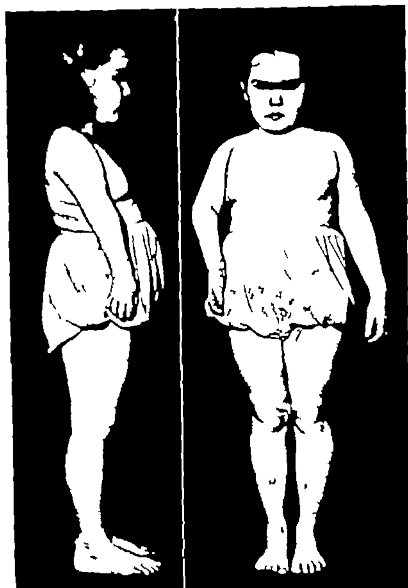
The internal rotation contracture results in the child holding the elbow away from the side in partial abduction in order to clear the arm from the trunk and enables one to diagnose this condition on the street. There is loss of humeral scapular rhythm with limited abduction most of which is really scapular elevation.

The humerus is frequently shortened out of all proportion to the muscular atrophy The acromion may be hooked but downward and elongated on x ray examination. Posterior subluxation or dislocation of the humeral head may occasionally be present.



Figure 361 Limited external rotation due to contracture in brachial palsy

Figure 362 Limitation of abduction and elevation despite presence of good deltoid power following birth type of brachial palsy



Figures 363 364 Whole arm involvement in brachial palsy forearm hand and wrist contractures.

Pronation or supination deformities may be present in the forearm depending on the muscle imbalance

Involvement of the deltoid biceps and external rotators at the shoulder is by far the most common muscle weakness observed. However it is not unusual on examining a late case to find good muscle power inhibited from maximum function by contracture

Treatment

Treatment cannot be based on the use of a position to prevent deformity twenty four hours a day Such therapy results in contracture in abduction and internal rotation. Periodic change into the corrective position is the guiding

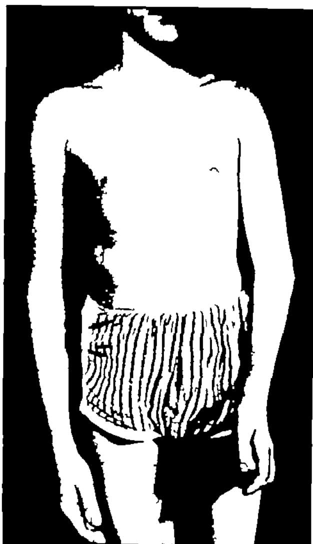


Figure 365 Upper arm involvement in brachial palsy Note shortening, tending to hold elbow away from trunk because of internal rotation contracture, flexed elbow but good hand.

philosophy of treatment. Neurological exploration and repair of the injury has not been justified—the proximal nature of the lesion and lack of suitable nerve sheath in young infants being major handicaps

The arm may be pinned in abduction and external rotation with a diaper while the infant is recumbent This should be for two to three hour periods only alternated with periods with the arm free. In addition at each diaper change the arm is passively abducted the elbow flexed the forearm supinated and the wrist dorsiflexed so that a full range of motion of all joints including fingers is gently carried out

More efficient corrective immobilization is carried out by a so-called Statue of Liberty splint which maintains abduction and external rotaton and in addition controls the forearm and hand in a manner which mere splinting cannot do The splint is not worn continuously It is alternated with periods entirely free and with periods of active and passive exercise The splint is usually needed for the first six months intermittently through day and night and perhaps for an additional six months at night only The child must be followed frequently

enough so that the earliest opportunity to emphasize scapulo-humeral rhythm in active use of the area is not missed About one half of the upper arm paralysis will respond to this regime with satisfactory shoulder function—but will be completely normal in only ten per cent

In late deformity the splint will not solve the clinical problem

The usual problem in late cases is to increase the range of external rotation and abduction If there is no bony deformity this may be accomplished in part by tenotomy of the subscapularis and pectoralis major muscles as described by Sever If active external rotation power is not present, it may be supplemented by a muscle transplant described by L'Episcopo In this procedure the teres major and often the latissimus dorsi are removed from their insertions and sutured under an osteoperiosteal flap on the posterior aspect of the humerus at the same level They are thereby converted from internal to external rotators. Abduction is frequently improved following these procedures as well

In the presence of functioning musculature, but fixed contracture, osteotomy of the humerus above insertion of the deltoid and rotating the distal fragment externally inside the periosteum may be indicated.

Improvement in cosmetic appearance as well as function can be expected from a correctly done operation for the right case

References

CLARK, L. P TAYLOR, A S AND PROUT T P A study of brachial birth palsy Am. J M Sc 130: 670 1905

FAIRBANK, H A. T Birth palsy subluxation of the shoulder joint in infants and young children Lancet 1: 1217 1913

L'EPISCOPO J B Tendon transplantation in obstetrical paralysis. Am. J Surg 25: 122 1931

L'EPISCOPO J B Restoration of muscle balance in the treatment of obstetrical paralysis. New York State J Med 39: 357 1939

SEVER, J W Obstetric paralysis its etiology pathology clinical aspects and treatment with a report of four hundred and seventy cases. Am J Dis. Child., 12: 541 1916

SEVER, J W The results of a new operation for obstetrical paralysis. Am J Ortho Surg 16: 248 1918

TAYLOR, E. S Brachial birth palsy and injuries of similar type in adults. Surg. Gynec & Obstet., 30: 494 1920

WICKSTROM J HASLAM, E. T AND HUTCHINSON R. The surgical management of residual deformities of the shoulder following birth injuries of the brachial plexus Jour Bone & Joint Surg 37A: 27 1955.

7

Affections of Bone

DEFECTIVE FORMATION OF BONE FROM CARTILAGE

ALBERT B FERGUSON SR., M.D

Chondrodysplasia and chondrodystrophy are terms which are often used loosely without exact definition. In this article they are interpreted as follows

Chondrodysplasia is defective formation of bone from cartilage resulting in relatively permanent defects in the bone structure

Chondrodystrophy is defective formation of bone from cartilage altering the size or contour of the bone but not resulting in permanent defects in the bone structure.

These definitions exclude transformation in formed bone—osteitis fibrosa of parathyroidism, benign giant cell tumor and benign aneurysmal bone cyst—and complete or partial absence of cartilage—hemivertebra incomplete segmentation of vertebrae (congenital fusion) and cleido-cranial dysostosis. Also excluded are effects on formed bone and on growth rate in affections of tissues about bone as in osteopsathyrosis myelomuscular disturbances and neurofibromatosis and conditions resulting from stress developed by resistance of soft tissues to elongation by the growing bones as in developmental or "congenital coxa vara, developmental genu valgum and developmental genu varum (Blount's disease Blount-Barber syndrome or osteochondrosis deformans tibiae)

CHONDRODYSTROPHIES

Chondrodystrophia fetalis (achondroplastic dwarfism) begins in fetal life and is often demonstrably familial. Diminished production of growth cartilage *with relatively prompt conversion of cartilage to bone* causes the growth disks to be grossly and roentgenographically thin The tubular bones are short and often somewhat distorted but adequately wide. The face is flat and the nose small and saddle-shaped but the vault of the skull is voluminous. The trunk is long relative to the short extremities this is an obvious difference from Morquio's disease in which the trunk is short. There is no effective treatment for the systemic condition Orthopedic treatment is rarely necessary but, if osteotomy



Figure 360 Achondroplastic dwarf with short extremities relative to the trunk

is contemplated for correction of deformity union of bone fragments follows the pattern for normal bones

Morquio's disease is a systemic disease in which chondrodystrophy is a feature It is mentioned here to point out that the dystrophy affects chiefly the formation of bone from vertebral and articular cartilage. Because metaphyseal bone formation is less disturbed the limbs are relatively long the trunk relatively short In spite of these characteristics an exact pattern of roentgenographic bone changes is not to be expected because the degree of chondrodystrophic change varies greatly from case to case and the dysfunctioning cartilage may also produce permanent defects in bone structure (chondrodysplasia), usually central type defects in the epiphyses and metaphyses



Figure 86 Upper extremities in chondrodystrophia fetalis (achondroplastic dwarf) Note relatively unaffected trunk

GARGOYLISM

Gargoylism (Hurler Pfaundler syndrome Dysostosis multiplex, Lipochondrodystrophy) is a disease of uncertain etiology which is sometimes familial. It begins to become apparent in the first year of life or soon thereafter with evidence of mental retardation infantilism and dwarfism. The skin becomes coarse and thick the corneas may be cloudy Pot-belly hepatomegaly and splenomegaly develop Hydrocephalus is frequent. The neck is usually short, dorsolumbar round back is present and mobility of the joints is impaired, especially in the spine and hands. Lipoid deposits are found in some cases (but not in others) in the liver spleen lymph nodes cornea brain and pituitary gland with enlargement of the sella turcica The central portion of the long bones, especially in the hands tends to be bulbous roentgenographically this suggests disturbance of accretion and resorption in formed bone rather than defective formation of bone cartilage. In the presence of such widespread disease chondrodystrophic changes appear relatively minor and incidental. They are of the type seen roentgenographically in Morquio s disease but are less marked. They consist of developmental dorsolumbar round back, and shortness of long bones with distorted metaphyses and underdeveloped epiphyses. The changes in the long bones are most evident in the distal portion of the upper extremity There is no effective treatment for the condition and no orthopedic treatment is advisable except the use of an extension frame or similar apparatus for inhibition or improvement of the round back.



Figure 308 Involvement of epiphyseal and articular cartilage at the hip in Morquio's Disease.



Figure 309 The hand in chondrodystrophy Note irregularity in metaphyseal area adjacent to the epiphyseal line and short broad phalanges



Figure 570 Chondrodystrophia multiflex congenita with multiple ossification centers in the epiphyses.

LOCAL CHONDRODYSTROPHIES

Chondrodystrophy may occur locally in a restricted area or in a single bone. Some examples follow.

Developmental dorso-lumbar round back. A vertebra near the dorso-lumbar junction, usually the twelfth dorsal or first lumbar, may be deficient in development at its anterior superior quadrant and its anterior surface. This may occur as an incidental feature in a systemic chondrodystrophy such as Morquio's disease but it is not infrequent as an isolated lesion. In the latter case deformity begins in infancy or shortly thereafter with development of rounded dorso-lumbar kyphos greater than the normal postural dorso lumbar round back of infancy. The deficiency of the vertebra is clearly evident and characteristic in the lateral roentgenogram. Two adjacent vertebrae may be affected. The de-



Figure 81 The spine in chondrodystrophy with increased anterior posterior diameter of the vertebra and bony deformity

formity tends to progress for a few years and then tends to decrease and improve. Reversion toward normal development may be aided by encouraging the prone and restricting the erect posture. When deformity is great recumbency in a posterior plaster shell or other apparatus maintaining moderate extension relative to the kyphos should be used to assure more prompt and adequate recovery from the developmental deficiency

DEVELOPMENTAL VALGUS OR VARUS OF A PHALANX

Developmental valgus (or varus) of a phalanx is characterised by moderate shortness of a phalanx with angulation of its distal articular plane transversely relative to its proximal articular plane. The middle phalanx of a finger is the usual site of the deformity but other phalanges and more than one phalanx may be affected. The condition is often demonstrably familial and gene controlled. In at least one family it is regarded as evidence of relationship It may be detectable at birth but usually is not recognised until bone development is well advanced The deformity does not reverse itself in the later stages of growth but the writer has not yet seen a case in which any treatment seemed advisable.

Delayed conversion of cartilage to bone is a descriptive term applicable to those rare cases in which the rate of formation of cartilage is normal but its conversion to bone is delayed The result is an unusual depth of the growth disk the writer has seen for example the apophyseal disk of each ulna measuring one inch in the axial diameter The condition has been observed only as a local chondrodystrophy The cases are too few to demonstrate familial occurrence It is disclosed only incidentally by roentgenogram since no symptoms or deformity develop No treatment is necessary

It should be noted here that pituitary gigantism exhibits increased rapidity of production of cartilage with relatively normal conversion to bone resulting in abnormally long bones with the growth disks roentgenographically normal and cretinism exhibits symmetrical slowing of production of cartilage and its conversion to bone so that the bones maintain normal roentgenographic appearance but take an abnormally long time to reach a given stage of development. These chondrodystrophic features of these diseases cause no noteworthy orthopedic problems.

Premature union of an epiphysis may cause shortness of one or more metacarpal or metatarsal bones in pseudohypoparathyroidism club foot, spina bifida, and in paralysis of the lower extremity occuring before the fourth birthday This is a form of local chondrodystrophy secondary to abnormality of adjacent soft tissues. In some instances it may be an inherited trait The shortness is seldom recognized before the eighth birthday The adjacent phalanx may also be affected. Treatment to correct the shortness is not necessary Partial premature union of an epiphysis following destruction of cartilage by injury or inflammation is not chondrodystrophy

CHONDRODYSPLASIAS

The important factors in the production of permanent defects of bone structure by defective conversion of cartilage to bone during growth are as follows

1 One or more of the many possible predisposing factors or exciting causes of such defective formation of bone must have been present. Among these are

   A. Normal probability that not all cell divisions will result normally

   B Increased probability for conversion defects when cartilage is older when it grows more rapidly or when it approaches completion of ossification. These factors make conversion defects rarer in infancy more common in adolescence some are not fully developed until after the pediatric age period They also do much to account for the facts that conversion defects are most common at the distal metaphysis of the femur which is the most rapidly growing metaphysis, that epiphyseal bone formed more slowly from articular cartilage is less subject to defects and that the portion of a long bone formed from fetal cartilage which is young and which grows little after it begins its conversion to bone is seldom the site of conversion defects.



Figure 5?3 Ollier's Disease Unilateral chondrodysplasia Note defects in structure of bone that is laid down.

Delayed conversion of cartilage to bone is a descriptive term applicable to those rare cases in which the rate of formation of cartilage is normal but its conversion to bone is delayed. The result is an unusual depth of the growth disk the writer has seen for example the apophyseal disk of each ulna measuring one inch in the axial diameter The condition has been observed only as a local chondrodystrophy *The cases are too few to demonstrate familial occurrence* It is disclosed only incidentally by roentgenogram since no symptoms or deformity develop No treatment is necessary

It should be noted here that pituitary gigantism exhibits increased rapidity of production of cartilage with relatively normal conversion to bone resulting in abnormally long bones with the growth disks roentgenographically normal and cretinism exhibits symmetrical slowing of production of cartilage and its conversion to bone so that the bones maintain normal roentgenographic appearance but take an abnormally long time to reach a given stage of development. These chondrodystrophic features of these diseases cause no noteworthy orthopedic problems

Premature union of an epiphysis may cause shortness of one or more metacarpal or metatarsal bones in pseudohypoparathyroidism club foot spina bifida, and in paralysis of the lower extremity occuring before the fourth birthday This is a form of local chondrodystrophy secondary to abnormality of adjacent soft tissues In some instances it may be an inherited trait. The shortness is seldom recognized before the eighth birthday The adjacent phalanx may also be affected. Treatment to correct the shortness is not necessary Partial premature union of an epiphysis following destruction of cartilage by injury or inflammation is not chondrodystrophy

CHONDRODYSPLASIAS

The important factors in the production of permanent defects of bone structure by defective conversion of cartilage to bone during growth are as follows

1 One or more of the many possible predisposing factors or exciting causes of such defective formation of bone must have been present Among these are

   A. Normal probability that not all cell divisions will result normally

   B Increased probability for conversion defects when cartilage is older, when it grows more rapidly or when it approaches completion of ossification. These factors make conversion defects rarer in infancy more common in adolescence some are not fully developed until after the pediatric age period They also do much to account for the facts that conversion defects are most common at the distal metaphysis of the femur which is the most rapidly growing metaphysis that emphyseal bone formed more slowly from articular cartilage is less subject to defects and that the portion of a long bone formed from fetal cartilage which is young and which grows little after it begins its conversion to bone is seldom the site of conversion defects.



Figure 57 Ollier Disease Unilateral chondrodysplasia Note defects in structure of bone that is laid down.

Delayed conversion of cartilage to bone is a descriptive term applicable to those rare cases in which the rate of formation of cartilage is normal but its conversion to bone is delayed The result is an unusual depth of the growth disk the writer has seen for example the apophyseal disk of each ulna measuring one inch in the axial diameter The condition has been observed only as a local chondrodystrophy The cases are too few to demonstrate familial occurrence It is disclosed only incidentally by roentgenogram since no symptoms or deformity develop No treatment is necessary

It should be noted here that pituitary gigantism exhibits increased rapidity of production of cartilage with relatively normal conversion to bone resulting in abnormally long bones with the growth disks roentgenographically normal and cretinism exhibits symmetrical slowing of production of cartilage and its conversion to bone so that the bones maintain normal roentgenographic appearance but take an abnormally long time to reach a given stage of development. These chondrodystrophic features of these diseases cause no noteworthy orthopedic problems

Premature union of an epiphysis may cause shortness of one or more metacarpal or metatarsal bones in pseudohypoparathyroidism club foot spina bifida, and in paralysis of the lower extremity occuring before the fourth birthday This is a form of local chondrodystrophy secondary to abnormality of adjacent soft tissues In some instances it may be an inherited trait. The shortness is seldom recognized before the eighth birthday The adjacent phalanx may also be affected. Treatment to correct the shortness is not necessary Partial premature union of an epiphysis following destruction of cartilage by injury or inflammation is not chondrodystrophy

CHONDRODYSPLASIAS

The important factors in the production of permanent defects of bone structure by defective conversion of cartilage to bone during growth are as follows

1 One or more of the many possible predisposing factors or exciting causes of such defective formation of bone must have been present. Among these are

   A. Normal probability that not all cell divisions will result normally

   B Increased probability for conversion defects when cartilage is older when it grows more rapidly or when it approaches completion of ossification. These factors make conversion defects rarer in infancy, more common in adolescence some are not fully developed until after the pediatric age period. They also do much to account for the facts that conversion defects are most common at the distal metaphysis of the femur which is the most rapidly growing metaphysis that epiphyseal bone formed more slowly from articular cartilage is less subject to defects and that the portion of a long bone formed from fetal cartilage which is young and which grows little after it begins its conversion to bone is seldom the site of conversion defects.



Figure 87 Ollier's Disease Unilateral chondrodysplasia Note defects in structure of bone that is laid down.

C Tension at the attachment of fibrous tissue to cartilage resulting from growth of the bone, especially rapid growth.

D Hemorrhage *or faulty development of vascular buds at the* area of conversion of cartilage to bone.

E Either hemorrhage or blebs adjacent to the conversion area.

F Localised injury of the conversion area.

G Familial tendencies or gene controlled factors affecting development as in multiple enchondromata of the hands or in diaphyseal aclasia.

H Endocrine disturbances affecting development as in cretinism para thyroidism, polyglandular dystrophy or Albright's disease.

I Affections of neighboring tissues which alter the nutrition of bone and cartilage as in Morquio s disease gargoylism and generalised neurofibromatosis

J Any other condition which disturbs the nutrition or functioning of cartilage or adjacent bone.

2. Material which is not bone is formed at the affected area. The material may be calcareous without bone structure or it may be cartilage or fibrous tissue Fibrous tissue may contain giant cells or cystic areas it may form one large cyst. The defect may contain a mixture of the materials mentioned *For most types of defect there is no constant relation between the* predisposing or causative factor and the character of the material in the defect.

3. The defective material is left behind in situ as the bone grows in length. The axial length *of the defect is thus determined by the time during* which defective conversion of cartilage to bone continues and the rate of growth in that time A defect which still extends to the cartilage at the time of examination is a continuous defect while one which has become separated from the cartilage by reversion to normal bone formation is an intermittent defect.

4 The defective material is not altered by the normal remodelling process in the shaft of a long bone nor is it subject to Wolff's law The extent to which the defect interferes with these processes depends chiefly upon whether the defect occupies or approaches the periphery of the bone and to a lesser degree upon the breadth and length of the defect and the number of defects.

5 The defective material does not tend to exhibit autonomous growth. A cyst may accumulate more fluid and become larger and more rounded with *resorption of adjacent bone* A *sufficiently broad defect may* increase slightly in greadth due to compression in the axial diameter by soft tissues resisting elongation by the growing bone. A cartilaginous defect may retain some of its physiological capacity to produce cartilage and bone and may express that capacity if stimulated by repeated motion or trauma. These are not autonomous growth Conversion defects practically never exhibit autonomous tumorous growth except in the few cases which develop secondary malignancy or the rare cases which develop large osteochondromata

6 Since conversion defects are often symptomless they may *attract notice*



Figure 573 The single exostosis—cartilage capped, remodelled at the base

only incidentally by roentgenographic examination late in life but they are formed during the growth period and are detectable in increasing frequency up to the end of adolescence

7 The great number of differences in appearance and character of conversion defects which may result from different combinations of the factors which have been mentioned is further augmented by variation in the number and distribution of the defects and by combinations of different defects The possibilities seem infinite

A few of these defects which are seen sufficiently often to appear to represent types will now be discussed. They are classified as peripheral submarginal or central defects at articular, metaphyseal or fetal cartilage.

PERIPHERAL CONVERSION DEFECTS

Single peripheral metaphyseal chondrodysplasia (exostosis osteoma, osteochondroma) A portion of cartilage derived from the epiphyseal disk becomes



Figure 8-4 Traumatic chondrodysplasia—the linear defect in the bone laid down from the epiphyseal line arose at the site of insertion of a Kirschner wire.

separated at the periosteal attachment The separated portion remains *in situ* in the periosteum and its radial distance from the center of the bone remains constant as it is left behind by growth at the metaphysis. If the separation begins sufficiently early in life the eventual site of the cartilage defect may be several inches from the metaphysis The defect preserves the underlying bone while adjacent bone is remodelled so that it soon appears as though there were a projection of bone with cartilage at its crest. The projection is directed away from the near metaphysis because resorption on the far surface of the exostosis is accelerated due to traction on the periosteum created by metaphyseal growth. The projection has the usual pencil type appearance when the defect became intermittent shortly after it began but a broader table type projection is de-



Figure 375 Multiple cartilaginous exostoses with short ulna and fibula and bowing of the radius.



Figure 376 See legend for Figure 375 above



Figure 377 Failure to remodel in defective formation of bone from cartilage Note persistent box-like metaphysis on left compared with remodelled metaphysis on right.

veloped when the duration of formation of the defect is longer The radial projection of the exostosis may be increased slightly by physiological activity of the cartilage cap producing some cartilage and bone If such activity causes the mass to be bulbous it may be called an osteoma if practically entirely bony an osteochondroma if bone and cartilage are obviously present. These distinctions and the diagnosis are usually made roentgenographically

No autonomous tumorous growth has been mentioned in the preceding description Usually there is none Occasionally the radial projection of the mass beyond its metaphyseal site is great enough to justify the suspicion that some autonomous growth has occurred and in rare instances an osteochondromatous mass several inches in diameter may eventually develop

These defects are innocuous and require no treatment except that a mass in an exposed position where it is subject to repeated irritation or traumatism from external sources or by movement of tissues over it may be removed by surgical excision at its base for the purpose of relieving discomfort or annoyance and as prophylaxis against secondary malignancy which eventually occurs with some frequency in such irritated lesions This treatment is definitely advisable when an exostosis on a rib is subject to friction by an adjacent rib in breathing.

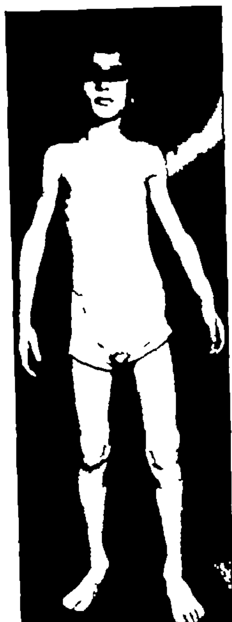


Figure 378 Patient with multiple cartilaginous exostoses involving the metaphyseal areas of the long bones.

MULTIPLE PERIPHERAL METAPHYSEAL CHONDRODYSPLASIA (DIAPHYSEAL ACLASIA)

Multiple peripheral metaphyseal chondrodysplasia (diaphyseal aclasia, multiple cartilaginous exostoses hereditary deforming chondrodysplasia, Ehrenfried's disease) is roughly symmetrical bilateral production of metaphyseal exostoses The long bones and ribs are principally involved but other bones may also be affected. Each exostosis develops and may be treated as described above for single exostosis. The condition is demonstrably familial and it affects males more often than females Central metaphyseal defects of various types may be detectable in some cases.

Selected cases exhibit various degrees of relative prominence of peripheral as opposed to central defects and of bilateral as opposed to unilateral distribution.



Figure 370 Epiphyseal osteochondroma involving medial condyle of the femur

They make it impossible to define exactly the limits of diaphyseal aclasia as compared to Ollier's disease.

UNILATERAL MULTIPLE PERIPHERAL AND CENTRAL METAPHYSEAL CHONDRODYSPLASIA (OLLIER'S DISEASE)

Unilateral multiple peripheral and central metaphyseal chondrodysplasia (unilateral dyschondroplasia Ollier's disease) is characterized by extensive unilateral involvement of metaphyses by an assortment of narrow and broad central and submarginal continuous defects (see below) mostly cartilaginous defects, with occasional fibrous defects and in some cases peripheral defects. In the older portion of the affected area formed before the metaphysis becomes completely

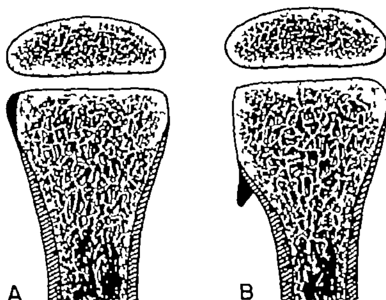


Figure 330 Diagram of the formation of exostosis which with its cartilage cap is not subjected to remodelling as is the uninvolved bone around it

involved axial striae may be seen representing thin lines of relatively normal ossification between defective areas The long bones of the lower extremity and the ilium are most commonly affected. Occasionally both sides of the body are affected and cases intermediate between Ollier s disease and diaphyseal aclasia are encountered. Cartilage in defects within the bone may undergo nodular calcification as life advances and in Ollier s disease this may begin to appear early even in childhood falsely suggesting a relationship to some form of calcinosis When the metaphysis is completely involved throughout its breadth the defective tissues are unable to overcome completely the resistance of soft tissues to elongation by the growing bone and growth in length becomes deficient This axial compression results in some lateral bulging of the defective tissue at the metaphysis the epiphyseal disk develops over the bulged tissue and the metaphysis thus becomes abnormally wide The same process may result in angular deformity when involvement is less complete toward one aspect of the metaphysis The defective formation of bone cannot be controlled. Angular deformity may be corrected by osteotomy with expectation of normal union of bone fragments Shortness of one leg may be compensated partially by stapling at epiphyseal disks in the opposite leg at an appropriate time or later a bone in the opposite leg may be shortened surgically by removal of a section

PERIPHERAL EPIPHYSEAL CHONDRODYSPLASIA

Peripheral epiphyseal chondrodysplasia produces separation of a cartilaginous conversion defect from articular cartilage at the site of attachment of fibrous tissue near the epiphyseal disk and the plaque of cartilage becomes increasingly separated from its area of origin by growth of both epiphyses at the joint until



Figure 331 Submarginal metaphyseal chondrodysplasia (Benign non-osteogenic fibroma) in the proximal femur bilaterally

its position is nearly opposite the center of articulation. Motion at the joint producing friction and stress at the cartilage defect stimulates its physiological capacity to grow and a mass one or two centimeters in diameter may result. Such cartilage is slow to ossify but it eventually does ossify centrally with cartilage usually persisting at its surface

The intermittent peripheral epiphyseal defect (sesamoidoma benign bone-forming tumor) thus forms a cartilaginous or bony mass in fibrous tissue at a joint suggesting an unusual sesamoid bone. The mass is rarely recognised before it ossifies As many as four or five such masses may be present at a joint. The knee or ankle is most commonly affected The condition is innocuous and may be ignored unless mechanical interference with motion makes surgical removal advisable

The continuous peripheral epiphyseal defect maintains continuity of cartilage between the defect and the epiphysis as the defect gradually becomes more displaced by growth of the two epiphyses The asymmetrical condylar mass thus formed distorts the joint in angular deformity which increases as the mass increases. Because of such deformity the patient is apt to be seen early before the cartilage at the defect ossifies. Roentgenogram then suggests the presence

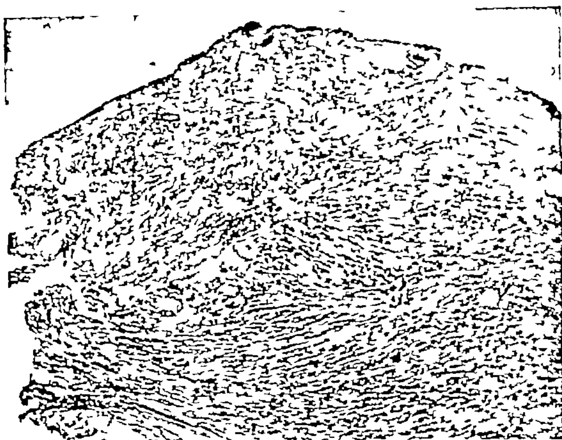


Figure 332 Fibrous tissue removed at biopsy from area of submarginal metaphyseal chondrodysplasia.

of a space occupying mass in the peripheral portion of the joint with corresponding angulation at the joint and underdevelopment of the adjacent aspects of the epiphyses to conform to the mass. No reactive phenomena are visible in adjacent bone or soft tissues. The knee is most commonly affected. When the patient is seen in this stage osteotomy through the shaft of the bone for correction of the angular deformity is not advisable because it does not restore normal conformation at the joint nor prevent further development of deformity On the other hand surgical treatment by paring off the adventitious cartilage sufficiently to approach normal conformation at the joint is feasible and proved satisfactory in the two cases thus treated under the writer's observation, the joint remained stable deformity did not increase appreciably and there was no stimulation of tumorous growth of the remaining cartilage. Later in life when physiological ossification has developed in the cartilage mass, treatment is less satisfactory At that time reshaping the distorted epiphysis is not feasible because a cartilaginous surface cannot be maintained and the condition may not justify such radical procedures as insertion of an internal prosthesis or fusion in corrected alignment restoration of alignment by osteotomy through the shaft of one of the bones is therefore the only treatment commonly justifiable and thus leaves a distorted joint operating in an abnormal plane Obviously



Figure 333 Ollier's dyschondroplasia with unilateral deformity

it is important to recognize and treat these patients early when the better result may be obtained.

SUBMARGINAL CONVERSION DEFECTS

Submarginal metaphyseal chondrodysplasia is defective formation of bone from cartilage situated within the bone but sufficiently near the periphery to be brought to the surface eventually as the defect is left behind by the growing metaphysis and the remodelling process causes the bone to become narrower. The defect may be recognised early when it is still entirely within the bone at the metaphyseal area or late when it is entirely at the surface of the remodelled narrow portion of the shaft but it is most characteristic in the intermediate stage when it is partly within the bone and partly at the surface with the perforation of the cortex parallel to the long axis of the bone but oblique to the flared



Figure 354 Roentgenogram of upper extremity in unilateral chondrodysplasia with involvement of long bones and metatarsals.

portion of the cortex which it perforates Submarginal defects may be broad enough to involve the peripheral or central areas or they may be accompanied by other defects in those areas. A thin line of cortex visible roentgenographically may be preserved over the defect by inhibition of remodelling and a thin calcareous wall may be formed about the defect by processes of accomodation in the adjacent bone structure The diameter of the defect varies with the breadth of the defective formation at different times during the period of continuity so that the sides of the defect may appear wavy or even scallopped.

BENIGN NON-OSTEOGENIC FIBROMA

Benign non-osteogenic fibroma is typically submarginal metaphyseal chondrodysplasia The writer has not yet seen a case in which there was histologic or roentgenographic evidence that the lesion was the site of autonomous tumor growth nor a case in which continued observation without surgical interference demonstrated increase in size of the lesion except by continued formation of defective material at the metaphysis The material at the defect is spindle cell fibrous tissue giant cells and foam cells may be present. The condition is innocuous, usually discovered incidentally by roentgenogram and requires no treatment although biopsy and more extensive surgery are often done If symptoms are present they can safely be assumed to be due to some other condition such as injury at the neighboring joint.

Calcareous submarginal chondrodysplasia is present when the contents of a submarginal defect are calcareous. The calcific material does not have



re 385 Central chondrodysplasia calcified in the adult (*From* Ferguson, A. B Jr. ed medullary defects in bone J Bone & Joint Surg., July 1947)



re 386 Calcified cartilage matrix removed from specimen illustrated in previous nogram (*From* Ferguson, A B Jr Calcified medullary defects in bone J Bone & urg July 194)

trabeculated or cortical bone structure and it appears amorphous in the roentgenogram in which it is readily visible because of its great density Such defects of moderate length are frequently observable in adolescence and later, especially in bones of the feet and in the fibula They may widen the cortical area peripherally or narrow the medullary canal endosteally or both depending upon their breadth and situation relative to the radius of the bone and the remodelled area of the bone They are innocuous and symptomless and require no treatment.

MELORRHEOSTOSIS

Melorrheostosis is calcareous submarginal chondrodysplasia developed in the metaphyseal and fetal areas along one aspect of a bone or succession of bones in a limb, tending to correspond with a somatic segment. It has been identified in childhood but usually does not attract attention until later when pain appears, motion becomes limited and calcification may develop in soft tissues about the joints Periosteum tends to adhere to the calcific mass and pain is probably the result of stress exerted by tissues attaching to the periosteum. The pain is relieved by rest. No other treatment is necessary Prognosis is good in that the condition is self limited and symptoms do not become marked.

Cartilaginous submarginal (and central) chondrodysplasia is characterised by the presence of cartilage throughout most or all of the defect. This character of the material is often not identifiable until the tissue is examined but it is suggested roentgenographically when the defect is eccentrically situated in the bone, irregularly rounded and not elongated axially and it is indicated in the adult when characteristic nodular calcification has developed in the cartilage of the defect. These defects are innocuous and symptomless unless they are in exposed situations where contusion or periosteal stress can excite physiological activity of the cartilage.

A small cartilaginous defect situated centrally in the medullary tissue where it does not deprive the bone of an appreciable amount of trabeculation is not detectable in childhood but becomes recognizable in the adult usually after age thirty, when nodular calcification occurs within it

A cartilaginous defect in the submarginal area brought to the surface of the cortex beneath the periosteum by the remodelling process may be too small to be detectable but may exhibit physiological activity of the cartilage in adolescence or later with formation of subperiosteal chondroma

A small cartilaginous conversion defect in fetal cartilage or early in metaphyseal growth is the probable source of the rare benign myxoma of bone of infancy

The broad central cartilaginous defect (benign central chondroma) may be encountered in childhood but usually does not attract attention until later A bone of the hands, feet or ribs is commonly affected The lesion is usually eccentric and the thin cortical line at its surface may undulate instead of presenting a smooth fusiform outline Its treatment is the same as for other broad central defects (*q v*) and it may not be distinguished from them until tissue is examined.

MULTIPLE ENCHONDROMATA

Multiple enchondromata of the hands is submarginal and central chondrodysplasia *which is apparently gene controlled. The feet may also be affected.* The defects are mostly submarginal and cartilaginous but tiny peripheral exostoses may be seen and some central fibrous or cystic defects may appear on tissue examination. The conversion defects are visible roentgenographically early in life (some may form from fetal cartilage) and they continue to form or be added to throughout most of the period of metaphyseal growth of the bones of the hands Physiological activity of the cartilage persists, probably because of the more or less constant use of the hands, and the cartilage masses tend to enlarge and may become quite big in relation to the size of the bones of origin. Individual masses which interfere with function or become tender due to repeated pressures may be removed surgically and a resulting cavity in the bone, if any may be filled with bone chips. Incomplete removal of a particular mass should be avoided if possible.

CENTRAL CONVERSION DEFECTS

Central chondrodysplasia develops conversion defects in the area of the bone which is not brought to the surface by the remodelling process. The defects may be *single or multiple, broad or narrow, long or short calcareous non* calcareous or mixed. When the breadth of the bone occupied by a defect or by a number of defects is sufficient, the operation of Wolff's law preserves enough bone about the defect to meet stresses normally applied to it and thus inhibits remodelling and presents the appearance of expansion' of the cortex. The cortex over such an area seen roentgenographically is thin and smoothly fusiform in shape except that rigid material (cartilage or calcareous material) in the defect may cause the outline to be wavy Such expansion is the hallmark of the large central defect.

Pyle's disease presents roentgenographic evidence of expansion toward the ends of the long bones (dumbbell bones) due to multiple non-calcareous defects which are individually too small to be visible. A similar condition may affect a single metaphyseal area symmetrically or eccentrically The condition is symptomless innocuous and requires no treatment.

Single broad central non-calcareous defects in metaphyseal chondrodysplasia (Osteitis fibrosa circumscripta, bone cyst central chondroma) presents roentgenographically a conspicuous area of deficient bone structure usually oval in shape *but with a transverse limit toward the epiphyseal* cartilage if it is continuous or only recently intermittent Expansion of the cortex is present to *the degree necessitated by the size and position of the lesion.* The position of the lesion in the long axis of the bone depends upon the length of the bone at the time the lesion began to form. It takes more than a year of defective bone formation to form a sizeable lesion and it may be years before it attracts attention The condition is therefore rarely encountered before the age of four *and frequency increases thereafter The material in the defect is* not positively identifiable before tissue examination but the roentgenogram suggests cartilage when the lesion is eccentric or its outline undulant and fibrous tissue if the lesion



Figure 85 Transverse defects laid down in the metaphyseal area adjacent to the epiphyseal line

is well separated from the metaphysis and symmetrical—the cyst tends to fracture before much separation from the metaphysis is achieved.

Broad central defects may be of mixed type containing some structureless calcareous material These may present a great variety of bizarre appearances on roentgenographic examination

A variation of the broad central defect occurs when the defect becomes intermittent shortly after its onset This produces only a thin plane of defective material which is left behind by the growing metaphysis. The thin defect is not visible roentgenographically until remodelling narrows the cortex to the diameter of the defect and then a thin transverse break in the continuity of the cortex is visible Transverse fracture occurs spontaneously at such a defect and if the defect does not extend completely across the bone it may be completed by trivial injury The writer believes that this type defect is the commonest cause of spontaneous fracture precisely in the transverse plane in childhood and early adolescence

Broad central calcareous defects in metaphyseal chondrodysplasia (marble bones osteopetrosis) exhibit roentgenographically a transverse dense area of structureless calcareous material toward the ends of the shafts. When the axial



Figure 388 Transverse "stress" type fracture in the tibia

extent of the defects is not great the condition is symptomless and requires no treatment. When the condition begins early in life (Albers-Schonberg's disease) the abnormal bone formation is generalized and may involve bone derived from fetal as well as from metaphyseal cartilage, little room remains for medullary tissue and anemia and activity of accessary hematopoietic organs develop blindness and deafness may result from faulty development of the skull. Predisposition to the disease is probably due to a gene-controlled Mendelian recessive character Blood chemistry and bone chemistry are essentially normal. Patients exhibiting the condition in infancy usually die young while those with later onset and sufficient medullary tissue in the bones have a good prognosis. The bones are subject to fracture which may be treated as similar fractures in normal bone. No other treatment is useful except medical supportive treatment to combat anemia

Multiple small central calcareous conversion defects of chondrodysplasia (osteopoikilosis) are usually systemically distributed in the epiphyses and,

toward the end of the growth period, the metaphyses. The defects are irregularly rounded and rarely over five millimeters in diameter They are symptomless and require no treatment. The prognosis is excellent although the defects are permanent.

Single small central calcareous conversion defects form single dense nodules, visible roentgenographically, which have often been described as bone whorls and as osteosclerosis The calcaneus, talus and proximal femur are common sites. The lesions are innocuous and should be ignored.

Small non calcareous central metaphyseal conversion defects may not deprive the bone of enough structure to be visible individually in the roentgenogram. They have been mentioned in association with other defects (Ollier's disease, Pyle's disease, *etc*) They may be visible individually if adjacent bone forms a calcareous limiting wall about them. Such a defect is indicated when a small circular area of deficient bone structure is outlined roentgenographically by a thin circular calcareous wall with no reaction in adjacent tissue. It is common in the femoral neck. It requires no treatment. It should be observed over a period of time to confirm its innocuous character rather than subjected to immediate biopsy

Defective bone formation may occur along the surface of epiphyses in Morquio's disease and in association with chondrodystrophy, spina bifida and other conditions but articular cartilage does not form non calcareous defects (visible within the epiphysis) as often as does metaphyseal cartilage. Because of slower growth, the epiphysis is less subject to defective formation and for the same reason the defect must continue for a longer time to achieve a size which is recognizable unless it is outlined by a calcareous ring as described above. Other single non-calcareous epiphyseal defects, if visible, are apt to occupy a considerable portion of the epiphysis and produce a bizarre effect. Lack of symptoms lack of reaction in adjacent tissues and the tendency of the sides of the defect to be straight and parallel to the direction of growth identify the defect in the roentgenogram. No treatment is necessary

The vertebrae are subject to non-calcareous conversion defects at their upper and lower surfaces (commonly called Schmorl's nodes) They may be detected by roentgenogram in mid-childhood as an early defect producing a shallow indentation of the ossified surface of the vertebra. If such a defect is followed through the growth period it will be observed to increase in depth exactly as bone is added to the vertebral surface by growth, the sides of the defect remaining perpendicular and the base parallel to the surface until late adolescence or later when it may become somewhat rounded by adjustment of adjacent bone to the defect. One or more such defects can be found in most spines in adolescence and later They are ordinarily symptomless and can be ignored but occasionally the developmental disturbance affects the cartilage more extensively and the underdeveloped disk may be thin due to deficiency of cartilage and nucleus pulposus. In the lumbar area where normal motion is considerable, such a lesion may produce local ache following activity because of the poor mechanical action of the joint.



Figure 339 Developing Schmorl's node



Figure 390 Same case as Figure 389 Two years later in same patient note that edges are straight bottom flat and that node has increased in depth and become well marked in the vertebra. A similar defect is beginning in the vertebra just above

METAPHYSEAL AFFECTIONS

MORQUIO'S DISEASE

The entity called Morquio's disease was first described by Morquio in 1929 as a "form of familial osseous dystrophy." Chondrodystrophy is characterized by an altered rate in the formation of bone from cartilage.

The characteristics of this disease include dwarf like stature, a marked kyphos usually centering at the dorso-lumbar junction, and involvement of the hips and other major joints in the changes of chondrodystrophy. These are noted clinically as enlargements and deformity. The bones of the skull and face are not involved and the intelligence is normal.

The disease has been recorded slightly more frequently in males but both sexes are affected. According to Fairbanks more than one member of a famiy is involved in about one-third of the cases. There is no apparent hereditary influence however.

Etiology

This wide-spread anomaly of bone structure and shape is unexplained on the basis of present knowledge.

Clinical Picture

The presence of Morquio's disease is ordinarily not recognized until after the child begins to walk. The disease is symptomless until the patient is older and secondary changes have developed in joints. The standing posture is very characteristic. The spine is in flexion with an accentuated kyphos at the mid spine area. The knees tend to be flexed and the feet pronated. Valgus at the knee is quite common. As the age of the patient progresses, these deformities tend to become more severe. The gait tends to be a bilateral Trendelenberg type resulting in waddling. This is due to the varus deformity at the hips.

The head may appear enlarged in relation to the short trunk and extremities, bus usually is normal for the chronological age in size and shape. The kyphos and round back results in a lordotic curve through the lumbar area usually quite short and accentuated. The spine shortening is out of proportion to that of the extremities. The hands may reach the knee level. There is limited extension of the spine and in older cases there may be some lateral elevation resulting in mild scoliosis.

In achondroplastic dwarfs the proximal portion of the extremities may be excessively short but this is not true in chondrodystrophy. The lower extremities in particular may have enlarged ends of the bones in the epiphyseal and metaphyseal areas. Such enlargements are more readily noted at the knee than elsewhere.

Motion at the hip joint which is usually involved in the disease is often limited and flexion contractures of mild degree present. There may be an inability to completely extend the knee. Cases have been described of excessive ligamentous laxity about the joints however. Enlargement of the interphalangeal joints is not common but may occur. The hand is broad with broad and blunt fingers.



Figure 301 The spine in Morquio's disease with abnormal shape, flattening and increased antero-posterior diameter of the vertebra.

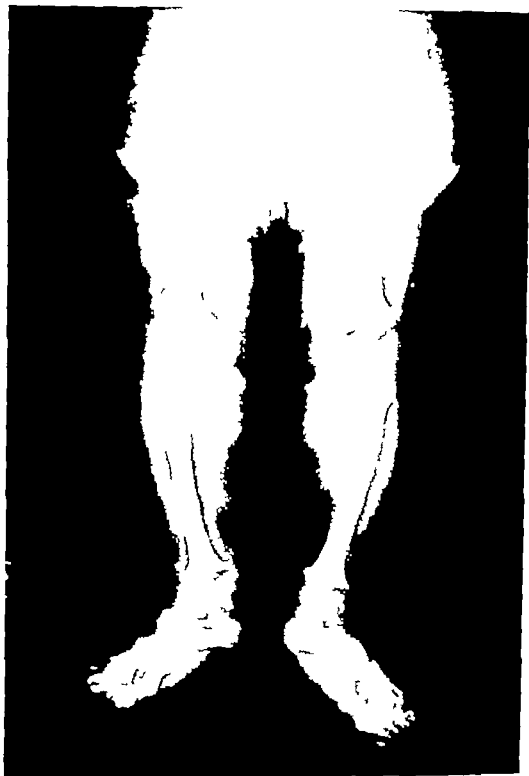


Figure 392 Short and broad bones with abnormal outline in chondrodystrophy



Figure 393 Hip joints, such as these in chondystrophic patient may later become the site of degenerative arthritis

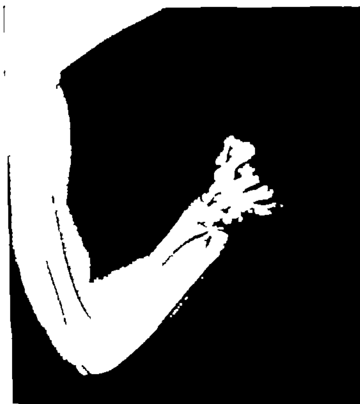


Figure 394 Forearm and hand with deformity and broadening of distal radius and shortened metacarpals.

Roentgen Findings

While the rate of formation of bone from cartilage may be altered there is no area of persistent failure of ossification as would be true in chondrodysplasia. Delayed growth of cartilage and failure of remodelling produces short and broad bones

The vertebrae have an increase in their transverse and antero-posterior diameters. The upper and lower surfaces are irregular and poorly defined. The anterior border may be much smaller than the posterior resulting in a general wedge shape. The wedging may appear to be more at the expense of either the upper or lower half rather than symmetrical however

At an area of kyphosis one vertebra may appear smaller and more irregularly wedged than the others and displaced posteriorly This vertebra is usually the first lumbar The epiphyseal lines of the long bones are widened, deepened and irregular The metaphyseal areas are broad out of proportion to the diaphyses. Not all epiphyses necessarily are involved.

The ribs are more horizontal than usual and may present an expansion at either end. The hips are involved, the changes becoming more evident with increasing age of the child The femoral heads show marked epiphyseal irregularity and flattening. The femoral necks are shortened broad and tend to a varus deformity According to Fairbanks, these changes are definitely progressive and may appear normal early in life only to develop later changes Other joints may or may not be involved.

The metacarpals and phalanges are short and expanded at their extremities. Ossification in the bones of the hand may be delayed and irregular in outline when it does appear

Differential Diagnosis

Achondroplasia is differentiated clinically and roentgenographically The spine is of normal height in achondroplasia the prominent buttocks and lumbar lordosis with a round back or kyphos are characteristic. The shortening of the limbs occurs principally in the proximal segments. Joint degenerative changes are not seen and genu valgus is not a feature The bone is affected with areas of failure to form bone rather than with slowness and irregularity of the rate of formation.

In dysostosis multiplex' or gargoylism the patient is mentally deficient the facial features coarsened and heavy the liver and spleen enlarged and the cornea cloudy

Tuberculosis and other causes of dorsal vertebral collapses and subsequent kyphos are readily distinguished clinically It should be remembered that chondrodystrophy of Morquio's type is a generalized disease and roentgenograms reveal this generalized involvement.

FAMILIAL METAPHYSEAL DYSPLASIA

A rare bone disease familial metaphyseal dysplasia was described in 1931 by Edwin Pyle. Its outstanding feature is a failure to remodel of bone laid down by growth from the epiphyseal line. The cases previously described in the



Figure 595 Familial metaphyseal dysplasia with failure of remodelling of bone laid down by the epiphyses. This disease indicates nicely the amount of growth from one epiphysis as compared to another

literature have had a familial incidence. The distribution is symmetrical with involvement of the metaphyseal areas of long bones. The wide flask shaped metaphyses may result in confusion with Gaucher's disease. Phalanges, meta carpals and metatarsals may also be involved

By roentgen examination the lack of involvement of bone of fetal or membranous origin is striking This results in pictures which show beautifully the relative percentages of growth from either end of a long bone. The area laid down by any one epiphyseal line has not been subject to remodelling. With the increased width there is a lack of normal development of bone cortex. The vertebra may be increased in the antero posterior diameter The pubis and ischii may be markedly widened Skull changes may be noted with a decrease in the transverse diameter at the base

The clinical picture is not striking The osseous lesions ordinarily are found incidentally on a routine x ray examination Valgus at the knee has resulted in its discovery in some children. Osteotomy may be necessary if the deformity justifies it. Treatment is ordinarily not otherwise indicated.

References

FELD H ET AL. Familial metaphyseal dysplasia. Radiology 65: 206, 1955
PYLE, E. Case of unusual bone development. J Bone & Joint Surg 13: 874 1931

SOLITARY BONE CYST

A solitary bone cyst is usually brought to the attention of the orthopedic surgeon by pathological fracture. Rarely they may be incidentally found on roentgen examination Although found in the adult, this lesion is peculiarly a disease of children and young adults It has been termed osteitis fibrosa but is not associated with the generalized skeletal changes found in hyperpara thyroidism, which is followed by polycystic lesions in the bones and bone atrophy

Silver reviewed the so called benign cyst of the bones in 1929 In ninety seven cases only eighteen were past the age in twenty years. The usual age group when bone cysts are discovered lies between six and fifteen years. In addition to the long bones, cysts occur in the calcaneous. The vast majority are in the humerus or femur, however There is a tendency for the lesion to predominate in males

Etiology

The underlying prime factor that causes the area immediately adjacent to an epiphyseal line to become productive of cyst tissue rather than bone is unknown Trauma and hemorrhage have been mentioned. There are two opposing theories of the mechanism of its production. The first assumes that the epiphyseal line in difficulty gives rise not to normal bone, but cyst tissue. This process may have already cased when first seen, and an area of normal bone may be visualized distal to the epiphyseal line. The walls of the cyst are never wider than the width of the epiphyseal line in solitary bone cyst.

An opposing theory notes that the mechanism of production could be an exaggeration of the resorptive phase when calcified cartilage is replaced by



Figure 300 The forearm and hand in Pyle's disease



Figure 397 Note small area of original fetal bone compared with unremodelled areas above and below in both tibia and fibula (familial metaphyseal dysplasia)

bone. This resorptive phase characterized by a proliferation of vessels and giant cell, according to Geschickter and Copeland may produce a loss of structure when exaggerated, resulting in the cyst.

The location is conspicuously that of areas of high rates of growth and extensive remodelling. What part trauma plays in the picture is still unknown.

Clinical Picture

Pain swelling and mild deformity may be complaints that cause the patient to seek medical attention. The pain is quite low-grade except in instances of acute fracture, and as a result may have existed for a considerable duration before being seen. There may have been no preceding symptoms. When present in areas such as the femoral neck, hemorrhage into the joint may initially result in confusion with entities that could cause pain and spasm at the hips.

The site of these complaints is characteristic The most common area involved is the proximal femur It is usually apparent on viewing the x ray film

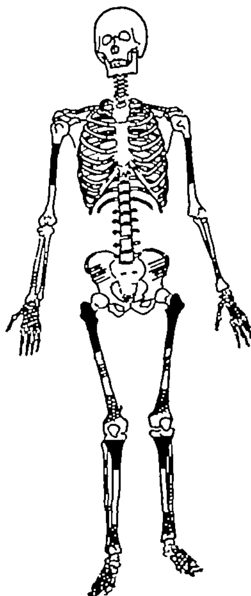


Figure 398 Diagram of distribution of solitary bone cyst. Solid black areas most frequent (*from* Geschickter and Copeland Tumors of Bone Lippincott, Philadelphia, 1949)

that either the trochanteric epiphysis or the capital femoral epiphyseal line is more closely related to the cyst. The second most common site is the proximal tibia. Other long bone sites may occasionally be involved The older the patient and the longer the duration of symptoms the further removed from the epiphyseal line the cyst may reasonably be found

So-called acute bone cysts may have a duration of but a few months and are regarded by Geschickter and Copeland as giant cell variants which may be related to giant cell tumor These cysts have not progressed down the shaft with growth but are situated close to the epiphyseal line Garceau and Gregory in tabulating the incidence of healing of the cyst in those cases with fracture found that it was fifteen per cent both in their own cases and in those in the literature.

Roentgen Findings

This lesion is not actually expansile although superficially it may appear so There is an area of loss of normal bone structure which is contained within thinned cortex. The actual width of the cyst will not be wider than the width of the epiphyseal line The cyst area has not been subject to remodelling, but this process has taken place distal to it. When the cyst is removed from the epiphyseal line remodelling has also taken place proximal to it. The remodelling of the normal bone results in the expanded appearance by contrast. When the width of the lesion is greater than that of the epiphyseal line from which it may have arisen, serious doubt of the diagnosis of solitary bone cyst should arise Lesions, such as syphilitic gumma and giant cell tumor should be considered

The juncture of the cyst with the normal *cortex is a symmetrical one* that is the normal cortex thins both on its exterior and interior surface to the width of the cortex overlying the lesion.

Pathology

The cyst wall on examination at operation may be greatly thinned and may fracture on slight pressure. There may be little or no lining but some areas of fibrous tissue are usually found. The cyst contents usually consist of fluid either yellow or reddened by recent hemorrhage Except in the region of recent fracture there is no indication of new bone formation going on exteriorly

In the areas of fibrous tissue lining the wall there is no evidence of fibrous tissue proliferation but there are occasionally osteoblasts and new bone The more central fibrous tissue if present is quite loose and myxomatous.

Evidence of old hemorrhages is sometimes noted In the areas of fresh hemorrhage a giant cell reaction may be present. Bone formation from cartilage is not seen and when there is osteoblastic activity it is taking place in fibrous tissue.

The *so-called giant cell variant of the solitary bone cyst* may have more tendency to be subcortical and most frequently is found in the region of the greater trochanter of the femur The tissue is featured by numerous giant cell areas close to the epiphyseal line in a stroma of considerable intercellular tissue and fibroblasts.

Treatment

The solitary bone cyst lends itself well to carefully performed surgery There has arisen a belief that fracture will heal cysts of this type but this belief is not borne out by experience Where the cyst is small with its long axis one-half inch or less the healing reaction about a fracture line extending about one quarter inch on either side of it may indeed ablate the cyst or render it of little consequence In the larger cysts such a reaction is not expected

When the cyst has suffered a recent fracture this healing reaction may be utilized by performing surgery during the phase of callus proliferation. Not all cysts must have surgery however

The optimum time for operation in a cyst which has rendered the bone mechanically inadequate arises when a centimeter or more of normal bone



Figure 309 Fracture in solitary bone cyst This will not result in healing of the cyst

Figure 400 Fibrous lining membrane from wall of simple bone cyst

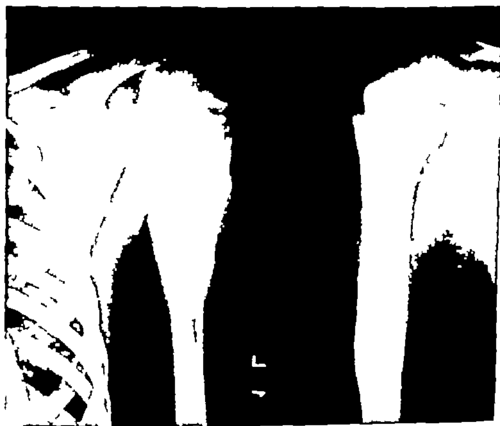


Figure 401 So called "immature state" of solitary bone cyst with epiphyseal line still directly adjacent to the cyst. Right post-operative view after bone grafting. (Courtesy of Elmer King M D)

has been laid down between the epiphyseal line and the cyst itself Attempts to ablate the cyst when there is no evidence that normal bone is being developed from the epiphyseal line may be followed by recurrence. Garceau and Gregory have emphasized the high recurrence rate in patients operated on below the age of ten.

The operation has two aims (1) to remove the cyst contents, (2) to stimulate a repair reaction which will result in bone being laid down in the cyst area At surgery the area involved is usually wider than the expected width of bone subject to remodelling and has a bluish cast. Normal bleeding bone should be visualized at both proximal and distal ends of the cyst cavity after curettement. There is occasionally a small quantity of fibrous tissue that can be obtained by curetting.

Bone grafts are inserted so that contact with the parent bone is made at either end of the cyst. If chips are used this is done in such a manner that large cystic spaces are not created which the bone would have difficulty in bridging. Nor is the cavity packed so tightly that the development of the repair reaction is impeded Autogenous bone is preferable but may be impractical in the presence of large cystic areas Where bone bank bone is used match stick grafts derived from ribs are inserted into the cyst cavity running longitudinally with good contact in normal cancellous bone areas

Carefully performed operative procedures in a patient in whom the cyst area is removed from the epiphyseal line by normal cancellous bone one centimeter or more in depth and in the older child, may be expected to yield good results

References

Geschickter, C T and Copeland M M Tumors of Bone. 3rd ed., J B Lippincott, Co Philadelphia 1949

Silver, D The so-called benign cyst of the bones. Am J Orthop Surg 9: 563 1912

Elmslie, R C Fibrocystic diseases of the bone. Brit. J Surg., 2: 17 1914.

Garceau G J and Gregory C. F Solitary unicameral bone cyst. J Bone & Joint Surg., 36A: 267 1954

Alldredge, R. H Localized fibrocystic disease of bone Results of treatment in one hundred and fifty-two cases. J Bone & Joint Surg., 24: 795 1942

Stewart M J and Hamel, H. A. Solitary bone cyst. Southern M J., 43: 927 1950

OSTEOMYELITIS

Although osteomyelitis occurs directly as following compound fractures, we are considering here principally the blood borne hematogenous infection of bone. This disease once occupied a most prominent position in texts as a difficult and dangerous entity requiring great skill in treatment The incidence of bone infection has been greatly reduced It still is possible, however, to regularly see unfortunate treatment situations in which the disease has been underestimated or not recognized. This is particularly true in the first three years of life It is primarily a disease of early childhood and is four times as frequent in boys as in girls

Osteomyelitis is most commonly caused by one of the pyogenic organisms. Green and Shannon have noted the higher incidence of streptoccal infections

apparently secondary to respiratory involvement in the first two years of life. The staphylococcus aureus rises in incidence as skin lesions liable to cause a bacteremia begin to occur in the ensuing decade

Etiology

The disease is most frequently secondary to a blood borne organism which tends to localize in the metaphysis of bones apparently due to a peculiarity of the vessels, *which with their anatomical loops tend to produce a relatively stagnant* situation.

The tendency to localize in the metaphysis is so strong that it frequently offers a helpful point in differential diagnosis Although the staphylococcus and streptococcus lead the list, infections with gram negative organisms occur and must be considered as a diagnostic possibility early in the disease when the organism may not yet be known. The pneumococcus, bicoli and typhoid bacillus will occasionally arise as the offending agent

Clinical Picture

The most usual history in these times of antibiotic agents is a primary infection respiratory skin abscess abscess of the teeth or intestinal infection which was treated apparently in adequate fashion However, a week or two weeks following the child may have a recurrence of fever lassitude and usually pain in an extremity becomes evident The primary infection may not have been sufficient for medical attention however and the child is first seen for his secondary infection.

If an infant, the tendency for the involved extremity to lie flaccidly may lead to confusion with diseases such as poliomyelitis If walking and the lower extremity are involved the limp may lead to similar confusion.

The most helpful diagnostic sign is the anatomic position of the tenderness which early in the disease will be maximal in the metaphyseal area. Late in the disease the possible spread of an abscess to involve the mid shaft results in more diffuse tenderness There is often local heat and swelling depending on the area involved The child frequently is in such agony with motion of the limb that any attempt to move it is resisted It is possible if one is careful not to press on a tender area to gain enough cooperation from the patient to carry the joints of the area through a range of motion and end in ruling out pathology in the joint itself

The child may be seriously ill with a septicemia in addition to his local disease The blood picture indicates an inflammatory response with a rise in polymorphonuclear leucocytes and increase in young forms. Rarely the osteomyelitis may be part of an overwhelming infection in infants without the usual systemic response

The antibiotic regime which the child has previously been subjected to may make diagnosis quite difficult. The bone may be carrying an abscess with a low grade inflammatory response development of physical signs may be slow and only a stubborn failure of the fever to subside from a low grade elevation to normal following treatment may provoke suspicion

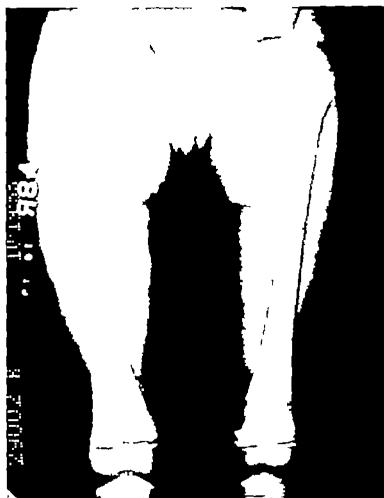


Figure 407 Osteomyelitis of proximal femoral metaphysis with involvement of the diaphysis sequestration of the shaft and wrapping of the diaphysis in reactive subperiosteal ossification (involucrum)

In infants osteomyelitis is still occasionally seen late in its development Such infections if in the region of the shoulder or hip may have broken through into the joint, and the proteolytic enzymes of the purulent material may have done irreparable damage to the cartilage of the epiphysis Phemister is responsible for calling our attention to the existence of these enzymes and the rapidity of their digestion of cartilage

Pathology

The metaphyseal abscess has many possible courses from its initial site It may subside with treatment and not extend A condensation of bone may form around it and the abscess become chronic This was more frequently seen before antibiotics and was known as Brodie s abscess The infection may spread through the cortex and elevate the periosteum forming a subperiosteal abscess A more massive development of the same trend results in the purulent material extending up and down the diaphysis and circumferentially around the bone The entire shaft or a portion of it may be cut off from its blood supply and a sequestrum may form. Such an avascular segment of bone becomes a complication in treatment. New bone of a coarse type may wall off the abscess and is called

involucrum. Pathological specimens have been obtained in which the original bone is encased with involucrum of similar form.

A less common course for the metaphyseal abscess is to rupture into the joint forming a pyogenic arthritis. Such a development is more common at the shoulder or hip where, depending on the location of the infection, little or no periosteum or fibrous septa may separate it from the joint space

While most commonly the localization of the disease is in the metaphyseal areas of long bones such as femur and tibia it should be remembered that other metaphyseal areas such as that of the os calcis have the same arterial configuration and are surprisingly common as sites of involvement.

Roentgen Picture

The x ray is very helpful and indeed most helpful early in the disease (the *first eleven days) when it is commonly stated that it is valueless.*

In order to be able to diagnose osteomyelitis at this stage, when it is most helpful to do so the examiner must be able to distinguish deep from superficial swelling. Deep swelling involves the area spoken of as of muscle density and having that anatomical configuration The area of the subcutaneous tissue is not involved. A superficial skin abscess on the other hand involves the subcutaneous tissue with increased vascular markings and the hazy loss of definition associated with inflammation The deep shadows in superficial cellulitis are not thickened or swollen. It is also necessary to remember that in the early stages, osteomyelitis is metaphyseal in location Thus a deep swelling of inflammatory type localized to the metaphysis is most helpful in recognizing osteomyelitis before changes of bone destruction have appeared

Later the bone destruction inflammatory swelling periosteal new bone (involucrum) and new bone distant from the lesion associated with stasis of the circulation make an obvious picture

Treatment

If the disease involves only a metaphyseal focus it is possible to treat and cure the disease with the correct antibiotic given a sufficiently long period of time to prevent recurrence and provided the limb is immobilized during that time

If there is a small subperiosteal abscess in addition a proportion of the cases may get by with antibiotics alone Most, however will need aspiration or drainage to avoid unnecessary complication such as interference with growth, the development of a sequestrum or a prolonged course.

Cases which have already developed beyond this point may need surgical intervention to save the child the limb the loss of an epiphyseal line or a joint or to secure a sequestrum and prevent the development of chronic osteomleyitis.

The judgement necessary to secure the maximum benefit for the child must be developed at the bedside not in a text but, in general unnecessary guttering and removal of bone is to be avoided. The healing powers of childhood are equal to restoring even a sequestration of the entire shaft. Cartilage of the epiphysis and epiphyseal line if damaged is damaged permanently, however

The general antibiotic approach is through wide spectrum drugs until the

Figures 403 404 The recognition of early osteomyelitis by deep metaphyseal swelling as compared to superficial cellulitis Top On the left the swelling involves superficial subcutaneous as well as deep tissues. On the right the swelling involves the deep soft tissues only Bottom On the left cellulitis area one week later On the right subperiosteal reaction can be seen eleven days later

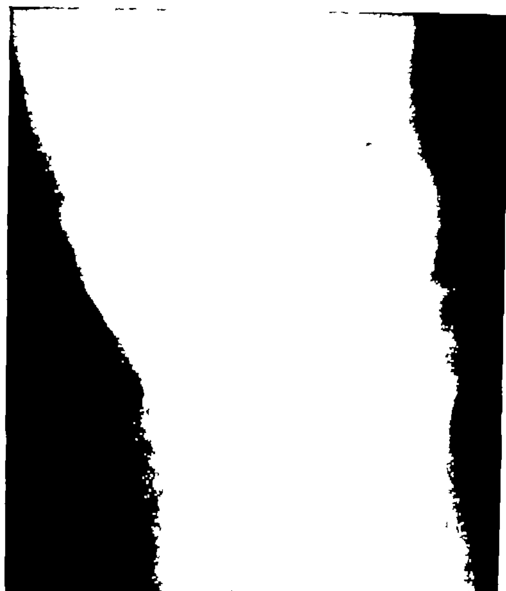


Figure 406 Osteomyelitis of proximal tibial metaphysis with bone abscess, destruction of adjacent cortex and periosteal reaction.

organism and the most useful drug are known. Dosage is high and kept up a minimum of three weeks and at least two weeks beyond the subsidence of fever and preferably also of the sedimentation rate The sedimentation rate may persist with a slight elevation for several months however if there has been extensive tissue destruction.

Rest is still good treatment for infection and in the presence of resistant organisms gives the natural antibody processes the greatest help in overcoming the disease Bed rest alone is not sufficient to truly rest the part This point has been found true many times A comfortable splint or bivalved cast insures real rest for the extremity involved.

References

BAKER, L. D AND SHANDS A R JR. Acute osteomyelitis with staphylococcus J A M A, 113: 2119 1939
BLANCHE, E. W Osteomyelitis in infants. J Bone & Joint Surg., 34A: 71 1952
BUCHANAN JOSEPH Acute hematogenous osteomyelitis American Academy Orthopedic Surgeon Instructional Course Lectures 9: 414 1952
BRIGHTMAN P AND RANTZ, L. A The clinical importance of coagulase-positive penicillin resistant staphylococcus aureus ? New England Med., 242: 353 1950
DICKSON F D The clinical diagnosis prognosis and treatment of acute hematogenous osteomyelitis. J A M A 127: 212 1945
GREEN W T AND SHANNON J C Osteomyelitis of infants A disease different from osteomyelitis in older children Arch Surg 32: 462 1936
WHITE, M AND DENNISON W M Acute hematogenous osteitis in childhood 34B: 608 1952

TIBIA VARA

There are three forms of tibia vara encountered in childhood. By far the commonest of these is tibia vara or bow legs associated with growth stress. Such a condition is commonly seen in orthopedic and pediatric practice. This form of tibia vara is associated with internal tibial torsion and is of mild degree It is present at birth—apparently a remnant of intrauterine position—and persists with stomach sleeping.

Deformity due to asymmetrical growth of the proximal tibial epiphyseal line with apparent osteochondritis (or Blount's disease) is occasionally seen and must be particularly considered in unilateral tibia vara Varus deformity of the tibia due to various forms of rickets and other growth disturbances of the epiphyseal line such as chondrodystrophy are now quite rare Where tibia vara occurs associated with generalized disease such as osteogenesis imperfecta it is usually an incidental part of a generalized deformity

Clinical Picture

Complaint of the deformity does not ordinarily take place until the child begins to walk and the rolling gait the wide-spaced knees and secondary pronation of the foot become evident Infants are seen in practice usually because the associated internal tibial torsion has resulted in turning inward of the foot to a sufficient degree to alarm the parents

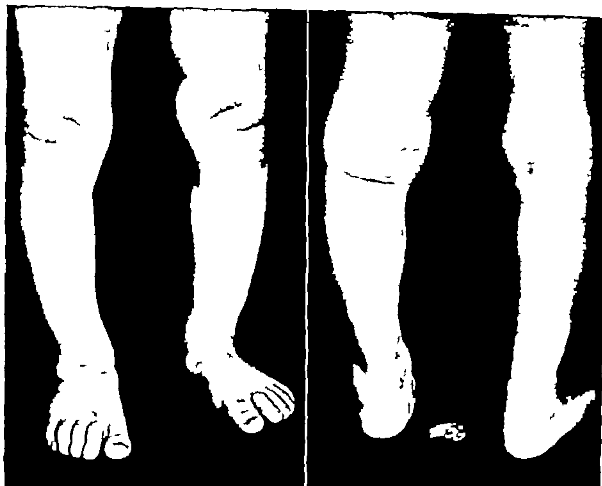
Infants with tibia vara due to growth stress are usually quite stocky and unusually active Those with Blount's disease do not exhibit evidence of generalized disease. However rachitic and other growth disturbances of the epiphyseal line usually reveal a generalized diminished rate of growth as well

Roentgen Findings

The findings on the x ray film are most helpful in differentiating the various causes of tibia vara.

Growth Stress

Here the angulation takes place in the proximal tibia. The epiphyseal line is normal There is a characteristic increase in width of the medial cortex most prominent at the junction of the proximal and middle third. The plane of the



Figures 406 407 Anterior and posterior views of patient with tibia vara. Note change of angle in proximal tibial area and secondary pronation of feet.

ankle joint faces slightly medially rather than parallel with the knee The deformity is equal bilaterally

Blount's Disease

There is diminished height of the proximal tibial epiphysis on its medial aspect. The epiphyseal line is narrowed and irregular on this side The metaphysis immediately below this area is irregularly ossified with beaking medially The diminished growth in this area results in angulation taking place proximally The deformity is often unilateral.

Rickets

The irregularity and widening of the epiphyseal line are evident at once The fraying of the metaphysis and its cup-like deformity are later changes along with generalized osteoporosis. The angulation of the tibia takes place at the junction of mid and distal third in contradistinction to that due to growth stress with maximum thickening of the medial cortex in this area. A similar angulation of the distal femur helps to contribute to the deformity



Figure 408 Weight bearing roentgenogram in tibia vara due to growth stress. Note medial facing of distal tibial surface and secondary pronation of feet.



Figure 400 Prominent beaking of medial metaphyses of both femora and tibiae Tendency to irregular ossification at proximal tibia on left is no longer present. One year later on right Severe though this appears one would expect these legs to straighten through growth with appropriate foot support.

Prognosis

Growth Stress

The tendency is for improvement of the deformity with weight bearing and growth Eventually at approximately age three to four years the deformity actually tends to pass over to genu valgus

Blount's Disease

The condition tends to subside spontaneously after a three to four year period with reconstitution of the epiphyseal line and filling in of the irregularly calcified medial metaphysis

Rickets

The response to antirachitic therapy is dependent on the type of rickets involved and is discussed elsewhere.



Figure 410 Blount's Disease At top irregular ossification and diminished height of metaphysis and epiphysis medially on right At bottom healing in previously afflicted area

Treatment

Tibia vara results in medial facing of the ankle joint and secondary pronation of the foot to get it flat on the floor In order to avoid this foot position it must be supported by a longitudinal arch pad. A one-eighth inch inner heel wedge is added to support the pad and stimulate medial growth. Following adequate alignment of the foot with the leg the child is followed, as growth itself tends to straighten the leg if the deformity is due to growth stress. Osteotomy of the tibia is not necessary

In Blount's disease the foot is similarly supported. The activity of the disease is followed by roentgenogram, and when the disease is quiescent and growth restored to normal surgical correction by osteotomy may be considered of the deformity is of sufficient degree Braces which are built with partial correction of the malalignment are sometimes used in an endeavor to prevent the development of severe deformity

The rachitic bow leg with deformity in the distal third of the tibia does not have the spontaneous tendency to correction that exists in angulation of the upper third. It is obviously poor judgement to attempt correction in the presence of active disease, but once the disease is controlled, osteotomy may be considered. This may involve correction of the femur as well, in order to get knee and ankle aligned in the same plane.

DIAPHYSEAL AFFECTIONS

ARACHNODACTYLY

Arachnodactyly is characterized by excessive length of the long bones. Accompanying abnormalities include ocular defects scoliosis, congenital heart disease and excessively loose ligaments. Arachnodactyly refers to the long, spider like fingers, but it is also known as Marfan's syndrome. Marfan in 1896 suggested the term "dolichostenomelia" emphasizing the long extremities.

Etiology

There is a strong hereditary and familial incidence apparent in this growth abnormality Occasional sporadic cases occur but more usual is the history of sibling and parental involvement.

Clinical Picture

The patient is often first seen because of noticeable deformity This may be scoliosis pronated feet or round back deformity The extremities appear wasted with very little subcutaneous fat The face is thin the patient spiritless and complains of a feeling of weakness to the examiner The ocular defect is usually displacement or subluxation of the lens although other defects have been reported. The feet are usually markedly pronated with occasional hammer toe deformities. The increased length of the extremity is more marked proximally than distally

The joints hyperextend in generally, but there may be contractures evident in fingers and toes Cardiac defects such as patent ductus or patent foramen



Figures 411 412 Diagnosis arachnodactyly Note long extremities and digits scoliosis and eye defect.

ovale may be present. Scoliosis is frequently accompanied by marked rib deformity and is primarily thoracic

Roentgen Picture

The excessive length of bone relative to the width is apparent The cortex is not well developed The elongation is evident in metacarpals and phalanges. Skeletal maturation is normal.



Figure 413 Femora in arachnodactyly

Figure 414 The roentgenogram of patient with arachnodactyly There is unusual length to long bones and phalanges and a mild scoliosis

Treatment

The treatment consists of support prevention and correction of the various deformities accompanying the primary condition Support for the feet and treatment of the eye condition are quite commonly necessary Generalized resistive forms of exercises are helpful in developing muscle power Care of the scoliosis falls under general principles described elsewhere

PROGRESSIVE DIAPHYSEAL DYSPLASIA

The rare disease progressive diaphyseal dysplasia is characterized by muscular wasting and the progressive laying down of subperiosteal new bone in the diaphyseal areas of long bones There are also mild neuromuscular disturbances The syndrome has also been termed Engleman's disease

Etiology

The cause is unknown Some observers have likened it to muscular dystrophy because of the progressive muscular weakness and familial tendency Instances of the disease have been found in relatives of the patient, who were unaware of any complaints

Clinical Picture

Males appear to be more commonly affected The disease is usually discovered in the four to ten year age group Easy fatigue or a gait abnormality usually bring the patient for examination This latter may consist of waddling difficulty in going up or down stairs or limp The patients are frequently under weight and show growth disturbance generally Pain is not a feature

The legs are thinned the reflexes may be hyperactive and there may be swelling and prominence of the diaphyseal areas of long bones Tenderness is not prominent The complete blood count serum calcium, phosphorous and alkaline phosphatase have not been noted to be remarkable

Roentgen Features

Neuhauser has emphasized the symmetrical skeletal distribution of the diaphyses of the long bones with thickening of the cortex both from the periosteal and endosteal side The epiphyses and metaphyses are normal and there is an abrupt termination of the lesion in extremities which are relatively long for the individual

Pathology

There is thickening of the periosteum with alteration of the cortex by bone resorption and accretion to result in a change to bone of cancellous type The marrow is frequently fibrous with mononuclear and hematopoietic foci. There is no evidence of the effect of an inflammatory agent Cartilage formation at the epiphyseal line and conversion to bone in the metaphysis are apparently normal.



Figure 416 Progressive diaphyseal dysplasia. The diaphysis is widened, the cortex is widened both from periosteal and endosteal side.

Treatment

Some individuals noted to have the disease are able to live full lives without disabling muscular weakness. Some have a progressive downhill course. Muscle exercises including those of resistive type to strengthen available musculature have been of some value. No medication has been proven to alter the course of the disease.

References

Neuhauser, E. B. D., Schwachman, H., Wittenborg, M., and Cohen, J.: Progressive diaphyseal dysplasia. Radiology 51: 11, 1948.

Riley, C. M., and Schwachman, H.: Unusual osseous disease with neurologic changes. Am. J. Dis. Child., 66: 150, 1943.



Figure 416 Involvement of diaphyses of the tibiae and fibulae in progressive diaphyseal dysplasia. Note symmetry of involvement.

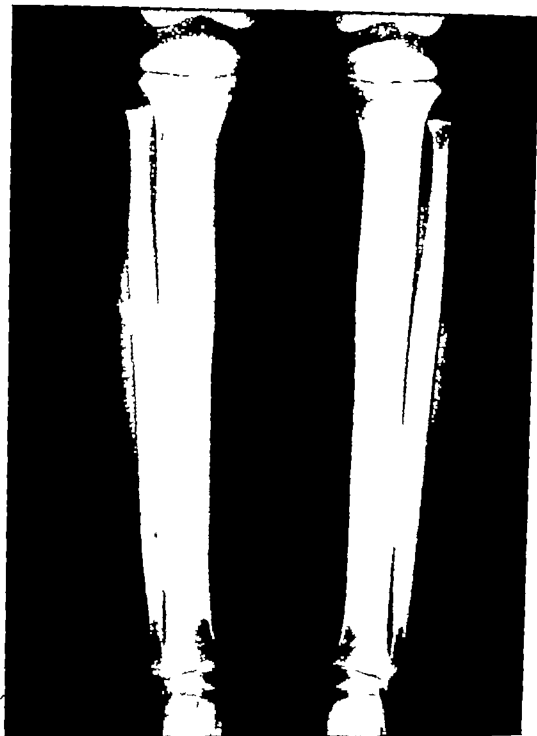


Figure 417 Same case as Figure 416. Three years later the cortex is beginning to have a mottled appearance

INFANTILE CORTICAL HYPEROSTOSIS

This is an infantile disease in which the diaphysis of long bones the mandible and the scapula are characteristically involved by the deposition of subperiosteal new bone It was first reported in the German literature by Roske in 1930, but was brought to American attention principally by Caffey (1939) whose name is sometimes given to this disease The mandibular swelling a principal sign, has led to an erroneous diagnosis of osteomyelitis in many cases The disease occurs in the first six months of life and is more common in the male

Clinical Picture

Swelling about the jaw without heat or redness leads to a suspicion of the diagnosis Swelling over the scapula is also especially characteristic, but not as frequent The infant is usually irritable and may have a low grade fever which runs a chronic course, if present Leukocytosis usually accompanies the fever

The swelling is often tender and asymmetrical The tibia humerus and clavicle are often involved in conjunction with the mandible and scapula. The small bones of the hands and feet are exempt along with the vertebrae

Roentgen Picture

Subperiosteal swelling occurs along the diaphysis starting usually centrally and then spreading throughout its length It may appear more marked on one side than on the other The edges of this new subperiosteal bone are smooth and laid upon the cortex in layers separated from it by a thin dark line. In later stages the subperiosteal new bone and the old cortex are not well delineated from each other, the area of new bone tending to become confluent. The new bone laid down in the scapular region is more mass-like and results in some confusion with tumor The lesion may be differentiated from osteomyelitis by the fact that the accompanying deep soft tissue swelling is diaphyseal rather than metaphyseal

Pathology

Biopsies have revealed the areas of subperiosteal bone formation but without any signs of active inflammation The marrow is often vascular and exhibits fibrosis with cellular activity in the form of osteoclasts Cultures of these lesions have been sterile

Treatment

The disease is self limited running up to ten or eleven months at the longest. It is not affected by antibiotics The swelling subsides and the bones appear normal eventually by roentgenogram. The disease is too widespread for osteomyelitis and does not exhibit bone destruction. Scurvy is inappropriate to the age group wherein this disease arises When the bone heals it does so by widening of the medullary cavity so that the cortex lies in the area of bony thickening. As the bone increases in width and length the developing bone catches up with the increased width created by the disease so that with sufficient growth the bone is reconstructed





Figures 418 419 Involvement of mandible in infantile cortical hyperostosis. (418 above) Lateral (419 below) Anterior view Clinically swelling due to this bony reaction may be mistaken for osteomyelitis.

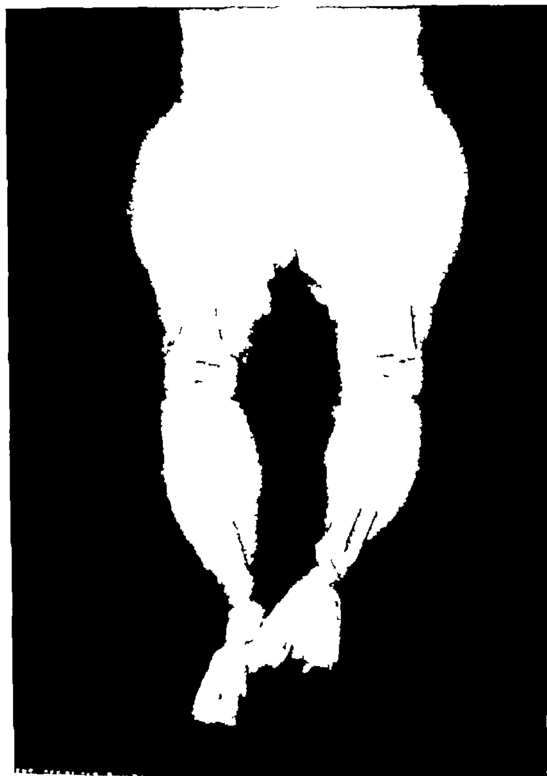


Figure [illegible] Early involvement of the tibia in Caffe's disease



Figure 6.1 Severe involvement with obliteration of bony architecture by bony reaction



Figure 423 Healing with widened shape maintained resulting in widened medullary cavity Eventually growth in length and width at the metaphysis will obliterate this evidence of disease

Reference

CAFFEY J AND SILVERMAN W A. Infantile cortical hyperostoses. Am. J Roentgenol, 54: 1 1945

OSTEOID OSTEOMA

Recognition of the osteoid osteoma lesion followed the publication in 1935 of a paper by Jaffe which brought out the clinical and pathological characteristics. It is seen as a cause of chronic bone pain in children, adolescents, and young adults. The nature of the lesion is still in doubt. Opinion supports a benign neoplasm, chronic infection and unusual bone repair Due to its unusual pathology and difficulty in classification it has occupied the attention of many writers with notable reviews published by Jaffe in 1945 and Sherman in 1947

Clinical Picture

The entity is more common in males with the majority of the cases lying in the age group between six and twenty years. It is rare in negroes. The two most common areas of involvement are the tibia and the femoral neck. Involvement of a vertebra is not unusual Other bones may be involved.

The usual patient is seen because of pain beginning insidiously and of many months duration The pain is accentuated at night, is localized, and may be relieved by aspirin. It characteristically has gradually increased in severity until medical advice is sought.

If in the lower extremity, examination frequently reveals an antalgic limp. There is usually quite severe atrophy of the soft tissues. A location close to a joint may cause limitation of motion. Superficial locations cause muscle spasm and limitation of motion and tenderness on palpation of the spine of the involved vertebra. There is no fever and no abnormality of routine blood studies.

Roentgen Examination

The characteristic picture is that of a radiolucent nidus surrounded by sclerosis. There may be a central calcified area within the nidus Demonstration of the radiolucent center may be quite difficult in the presence of excessive sclerosis. With a cortical location the periosteum is often raised with considerable thickening of the bone Lesions in an early stage of development with minimal bony reaction may easily be missed.

Pathology

There is a vascular fibrous tissue stroma which may be quite cellular Occasional giant cells may be found. Most characteristic is an irregular deposition of osteoid tissue scattered throughout the nidus without a pattern. Some of the osteoid trabeculae may become bone. Surrounding the nidus are areas of hypertrophic bone often with intervening fibrous tissue rather than marrow between the layers. There is no evidence of acute or chronic inflammation.

Etiology

Aside from occasional cultured organisms from the group that commonly are thought of as contaminants there is no evidence that osteoid osteoma is an in-

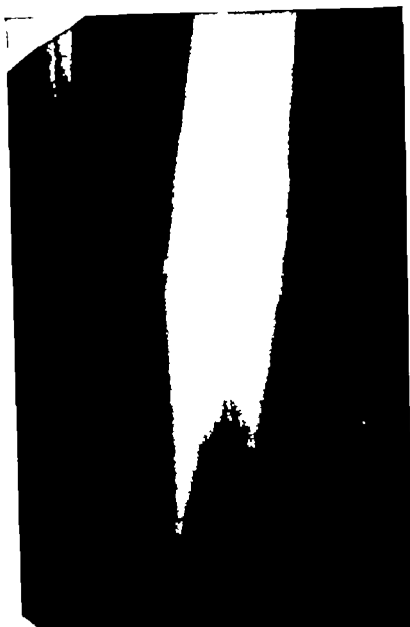


Figure 423 Subperiosteal and endosteal ossification in diaphysis of tibia in osteoid osteoma. The area of the nidus is well delineated

fection. There is no clinical response to infection and no pathological evidence of such. Jaffe supports the theory that these are benign neoplasms but there is no evidence that this "tumor" grows The nidus is always small

The lesion may represent repair following trauma which for some reason can not be carried to its full conclusion. This excites the outpouring of further bony calcification to wall off the defect. The repair under difficulty may be caused by a vascular lesion

Reference

Caffey J and Silverman W A. Infantile cortical hyperostoses. Am. J Roentgenol, 54: 1 1945

OSTEOID OSTEOMA

Recognition of the osteoid osteoma lesion followed the publication in 1935 of a paper by Jaffe which brought out the clinical and pathological characteristics. It is seen as a cause of chronic bone pain in children, adolescents and young adults. The nature of the lesion is still in doubt. Opinion supports a benign neoplasm, chronic infection and unusual bone repair Due to its unusual pathology and difficulty in classification it has occupied the attention of many writers with notable reviews published by Jaffe in 1945 and Sherman in 1947

Clinical Picture

The entity is more common in males with the majority of the cases lying in the age group between six and twenty years It is rare in negroes. The two most common areas of involvement are the tibia and the femoral neck. Involvement of a vertebra is not unusual Other bones may be involved.

The usual patient is seen because of pain beginning insidiously and of many months duration. The pain is accentuated at night, is localized and may be relieved by aspirin It characteristically has gradually increased in severity until medical advice is sought.

If in the lower extremity, examination frequently reveals an antalgic limp. There is usually quite severe atrophy of the soft tissues. A location close to a joint may cause limitation of motion Superficial locations cause muscle spasm and limitation of motion, and tenderness on palpation of the spine of the involved vertebra. There is no fever and no abnormality of routine blood studies.

Roentgen Examination

The characteristic picture is that of a radiolucent nidus surrounded by sclerosis. There may be a central calcified area within the nidus. Demonstration of the radiolucent center may be quite difficult in the presence of excessive sclerosis With a cortical location the periosteum is often raised with considerable thickening of the bone Lesions in an early stage of development with minimal bony reaction may easily be missed.

Pathology

There is a vascular fibrous tissue stroma which may be quite cellular Occasional giant cells may be found. Most characteristic is an irregular deposition of osteoid tissue scattered throughout the nidus without a pattern. Some of the osteoid trabeculae may become bone Surrounding the nidus are areas of hypertrophic bone often with intervening fibrous tissue rather than marrow between the layers There is no evidence of acute or chronic inflammation.

Etiology

Aside from occasional cultured organisms from the group that commonly are thought of as contaminants there is no evidence that osteoid osteoma is an in

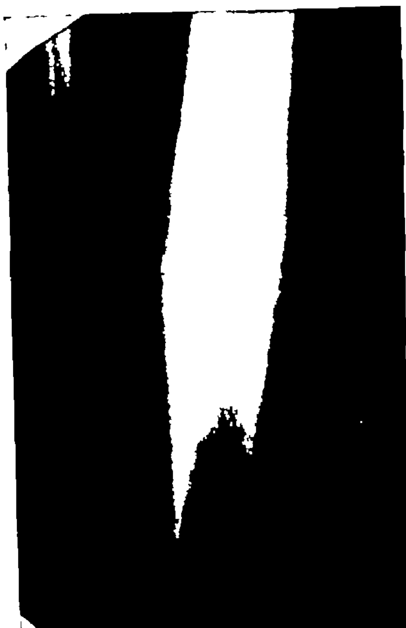


Figure 4.3 Subperiosteal and endosteal ossification in diaphysis of tibia in osteoid osteoma. The area of the nidus is well delineated.

fection There is no clinical response to infection and no pathological evidence of such Jaffe supports the theory that these are benign neoplasms but there is no evidence that this "tumor" grows The nidus is always small

The lesion may represent repair following trauma which for some reason can not be carried to its full conclusion This excites the outpouring of further bony calcification to wall off the defect The repair under difficulty may be caused by a vascular lesion

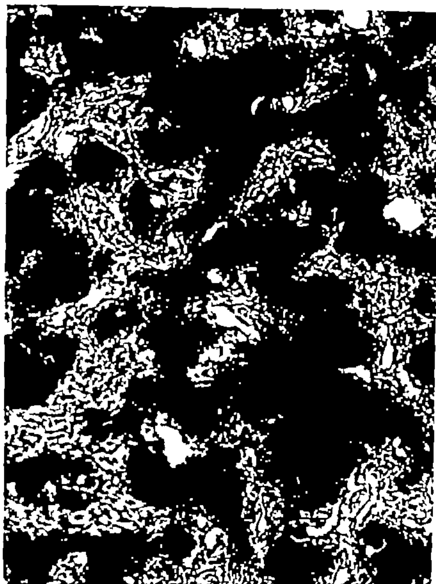


Figure 424 Osteoid formation from osteoid osteoma

Treatment

The mature course of osteoid osteoma appears to be chronic persistence of the lesion. Some authors have reported cases in which after a period of years the nidus and pain cleared although bony sclerosis was still evident.

The response to surgery which removes the nidus is dramatic and satisfactory It is appropriate to insure removal of the essential part of the lesion—the nidus. This is done by inserting drills above and below the area presumed to contain the lesion An x ray is then taken in the operating room On demonstrating that the nidus lies between the drills it is removed en bloc.

References

JAFFE H L. Osteoid osteoma A benign osteoblastic tumor composed of osteoid and atypical bone Arch. Surg 31: 709 1935
JAFFE, H L. Osteoid-osteoma of bone Radiology 45: 319 1945
SHERMAN M S Osteoid-osteoma (review of the literature and report of thirty cases) J Bone & Joint Surg., 29: 918 1947
WALLACE, G T Some surgical aspects of osteoid-osteoma J Bone & Joint Surg., 29: 777 1947

PSEUDARTHROSES IN CHILDHOOD

There is a varied background to pseudarthroses in childhood. The congenital type is rare by comparison with other anomalies. It is noted at birth or shortly thereafter and is most frequent in the mid or distal third of the tibia or fibula. They have also been found in the femur clavicle, humerus, ulna and first rib

Strangely some pseudarthroses appear to develop with gradual narrowing of the bone The medullary cavity becomes sclerosed and a fracture either spontaneous or induced occurs. The end result of both types appears the same The area of the pseudarthrosis consists of the meeting place of two narrowed bony structures both of which have lost their medullary cavity Green and Rudo reported the finding of neurofibroma at this site This has been confirmed by similar findings in other cases. However, in some cases little is noted other than a binding structure of dense connective tissue about the pseudarthrosis

The area of pseudarthrosis is sometimes bound by a bulbous mass consisting of cartilage and fibrous tissue in an irregular distribution without marked narrowing of the bone. In such cases there may be adjacent cystic changes in the bone.

Considerable overlapping often ocurs which masks the actual loss of substance in the tibia and results in considerable shortening of the affected leg.

MacFarland feels that the bending stress in these angulated tibias stimulates resorption over accretion with a fatigue fracture occuring in defective bone. His by pass graft attacks the lesion on the basis of this theory

Treatment

The history of treatment includes many well thought out and noble attempts to secure union With some exceptions many of these attempts have been quite discouraging.

J R Moore has described a procedure in which the united fragments of the tibia are each held by two transfixing pins and placed in an Abbott leg lengthening apparatus An os novum type of graft is raised on the opposite tibia and used eighteen days later to bridge the defect along with a smaller fibular graft. The pertinent points in technique as detailed by Moore are that (1) the bone grafts must overlap the fragments two inches or more, (2) fixation with four pins is essential (3) the graft is held in position until a well defined medullary cavity has appeared (4) the graft should not be delayed more than eighteen days

MacFarland has used two procedures One is a double onlay graft later also described by Boyd (1941) The second is a by pass graft technique in which a



Figure 425 Congenital anterior angulation of the tibia possibly preliminary to pseudarthrosis

tibial cortical and cancellous graft from the opposite leg is inserted into the proximal and distal tibial fragments. The graft is posterior and does not directly attack the pseudarthrosis It is so placed that the stress of use will be transmitted through it causing hypertrophy

MacFarland has favored the by pass technique rather than the double onlay graft, as he feels it is more certain to produce a good result.

Charnley has used intramedullary fixation including tibia and os calcis along with grafting but without lengthening with success.

Whatever method is used, the chances of success are slim unless the greatest care is used to insure that no fundamental principle in the treatment of non-union is neglected. This includes elimination of infection adequate fixation, good soft tissue coverage wide bone contact and good general health of the patient and placement so the graft will be functional. Once union is secured, regrafting should not be delayed if the bone shows a tendency to narrow in any portion. Gradually increased protected weight bearing will help to develop the bone

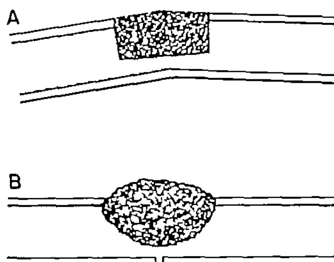


Figure 426 Diagram of delayed osteotomy used in correcting angulations where bone healing is suspect. (A) Initial operation dividing one half of involved bone and leaving mass of bone chips. (B) Second operation with division of second half of bone when callus has formed at first area.

Figure 427 Pseudarthrosis of tibia and fibula with narrowing of bone progressively toward involved area.

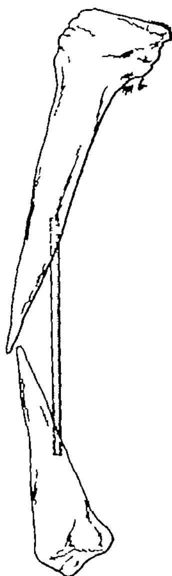


Figure 448 Diagram of by-pass, cortical graft of McFarland Bone chips may also be used to fill in interval between graft and parent bone.

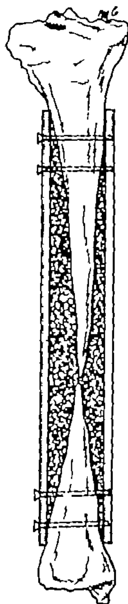


Figure 449 Double onlay grafts for pseudarthrosis with double fixation of both proximal and distal end bone chips filling in area of bone narrowing (Moore and Boyd with modifications)

References

GREEN W T AND RUDO W Pseudarthrosis and neurofibromatosis. Arch Surg 46: 639 1943.

MACFARLAND B Pseudarthrosis of the tibia in childhood J Bone & Joint Surg., 33B: 36 1951

NEUROFIBROMATOSIS

Manifestations of neurofibromatosis are seen in childhood as the cause of some difficult and bizarre orthopedic problems The presence of cafe au lait spots as a skin pigmentation and skeletal and joint changes are seen as an entity in a variety of conditions

Von Recklinghausen originally described the relationship between peripheral nerves and the subcutaneous nodules which are typical of the disease in the adult. This advanced form of the disease described in 1882 has been supplemented by the childhood manifestations in which tumors of the skin and subcutaneous tissue are seldom evident visually, but may be palpable.

The particularly evident forms of the disease, all forming difficult treatment problems are congenital pseudoarthrosis scoliosis localized and generalized hypertrophy of a limb with resultant length inequality and sarcoma associated with the disease

In the spine the disease produces a scoliosis which is sharp, acute and short in its primary curve Such a scoliosis tends to be rigidly progressive. It may be associated with chest masses and widened neural foramina with tumor also in the neural canal

In localized areas of hypertrophy the skin pigmentation, palpable nodules enlargement of the part and erythema are all seen. Such a process may involve an entire leg giving rise to a picture very much like elephantiasis. In a growing child such a process produces great inequality of leg length with overgrowth of the affected side

Green and Rudo reported the finding of neurofibroma at the site of a congenital pseudarthrosis. While this disease may not be the cause of all such pseudarthroses, it appears to be involved in many and renders the problem of securing union very difficult.

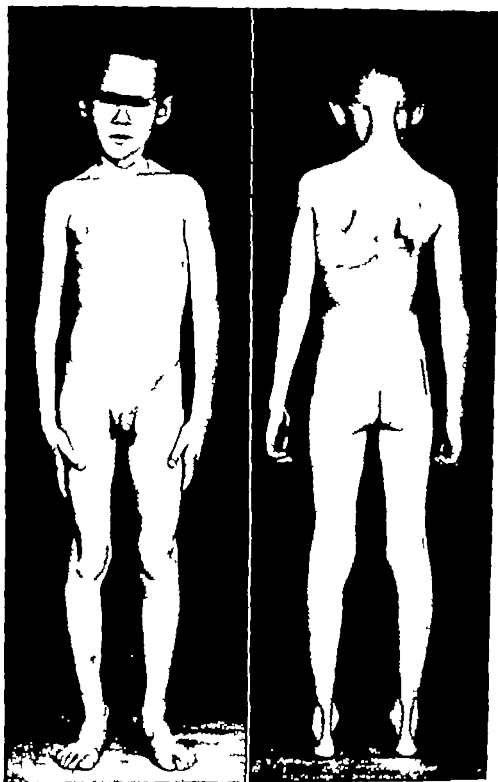
Sarcomas associated with the disease are found often contiguous with other neurofibromatous tissue. Such sarcomas are difficult to excise as they ramify through and around major nerve plexuses and appear usually as a neurofibrosarcoma under the microscope. Bone cysts in localized subperiosteal areas have been described The treatment of these various lesions is highly individualized and the general principles have been described elsewhere.

References

McCARROLL, H R. Clinical manifestations of congenital neurofibromatosis. J Bone & Joint Surg 32A: 601 1950

GREEN W T AND RUDO W Pseudarthrosis and neurofibromatosis. Arch. Surg., 46: 639 1943

MOORE, B H Some orthopedic relationships of neurofibromatosis. J Bone & Joint Surg 23: 109 1941



Figures 430, 431 Patient with neurofibromatosis involving spine There is a mild structural scoliosis of the dorsal spine and cafe-au-lait spots of skin pigmentation

METABOLIC BONE AFFECTIONS

GAUCHER'S DISEASE

Gaucher's disease has been known since its description in 1882. It was thought originally to be a neoplasm. Cushing and Stout in 1926 reported a case involving the hip. There was marked collapse of the proximal femur and acetabulum. This case in a woman of thirty three was the first recognition of bone involvement in association with the disease. It represents a rare familial disorder of lipoid metabolism in which kerosin is deposited in the reticulum cells of the reticulo-endothelial system. These cells, which have a typical appearance, are called Gaucher cells and may appear in the spleen, liver, lymph nodes and bone marrow. It is most common in Jewish patients. Symptoms are usually noted in childhood and adolescence, but the disease is chronic and only slowly progressive.

The most common area of bone involvement is the lower femur (about two-thirds of the cases) with the hip second in frequency.

According to Schein and Arkin the skeletal changes are due to infiltration and replacement of the bone trabeculae by kerosin containing reticulum cells. This infiltration results in linear radio-lucent areas giving the bone a characteristic mottled appearance. The cortex tends to become thinned and the bone slightly but not markedly expanded in the metaphyseal area. The expansion is fusiform rather than lobular. Secondary changes may occur particularly at the hip where the femoral head may collapse or undergo aseptic necrosis. Pathological fracture of the femoral neck and a tendency to coxa vara also occur. Vertebral involvement and collapse are also known.

These bone changes in a patient with splenomegaly, hepatomegaly, secondary anemia, hemorrhages and brown pigmentation of the skin result in the diagnosis of Gaucher's disease. The suspicion can be confirmed by bone marrow biopsy.

Treatment

The care of these skeletal lesions consists of measures to relieve pain and prevent deformity. Bed rest is indicated in periods of acute symptoms. The hip joint and femoral neck lesions must frequently be kept from the hazards of weight bearing and subsequent deformity by non weight bearing splints or when indicated by plaster spicas. In the carefully followed case regeneration of the femoral head and development of more normal bone structure has been observed.

References

CUSHING, E. H. AND STOUT, A. P. Gaucher's disease with report of a case showing bone disintegration and joint involvement. Arch. Surg., 12: 539, 1926.

GAUCHER, P. C. E. De l'epithelioma primitif de la rate hypertrophie de la rate sans leucemie. These de Paris, 1882.

MILCH, H. AND POMERANZ, W. Bone changes in Gaucher's splenomegaly. Arch. Int. Med. 61: 793, 1938.

SCHEIN, A. J. AND ARKIN, A. M. Hip joint involvement in Gaucher's disease. J. Bone & Joint Surg. 24: 396, 1942.



Figure 458 Involvement of the distal femur in Gaucher's disease

EOSINOPHILIC GRANULOMA

When confronted with a cystic type lesion of bone one of the considered diagnoses is that of eosinophilic granuloma. Such a lesion totally destroys the central area that it occupies It is most common in the skull but often is seen as an isolated lesion of the long bones. Such a lesion may be a forerunner of the similar lesions of the skeleton and may then be combined with visceral lesions to form the entity known as Hand-Schüller Christian syndrome. It is much more common in children than in adults.



Figure ‚33 The femoral neck can be weakened to the point where a coxa vara deformity gradually develops in Gaucher's disease



Figure 5 Aseptic necrosis of the femoral head as a complication in Gaucher's disease



Figure 435 Granuloma involving lateral portion of lower eyelid (eosinophilic granuloma)



Figure 436 The same patient as in Figure 435 four weeks following roentgen therapy to the granuloma.

Pathology

Farber has studied this disease extensively and concluded that the so-called eosinophilic granuloma represents one variant of a more basic disease process grouped under the general heading of xanthoma. The lesion is a granuloma which appears gelatinous and reddish yellow on gross examination. Microscopically there are numerous large mononuclear phagocytes with granular to finely vacuolated cytoplasm. A prominent feature consists of numerous eosinophiles. The relative prominence of eosinophiles may be evident clumps or they may be scattered diffusely. The more mature granulomas are featured by large vacuolated mononuclear cells rather than eosinophiles.

Roentgen Findings

The contour of the individual lesions is variable in outline but tends to be either oval or round. They are totally destructive, that is, no bone remnants are contained within the outlined area of the lesion. The cortex is not expanded but may be destroyed. The edge of the lesion usually appears well outlined and benign, but in some lesions particularly in the skull it is minutely spiculated. The reaction about the lesion in surrounding bone consists of minimal increased deposition of bone rather than marked sclerosis.

The lesions are most common in the skull and ribs. Flat bones are predominantly involved. When the bones of the extremity are involved the area closest to the trunk is the more usual site. Involvement of the distal end of the extremities is virtually unknown. There is no particular predominant site within the long bones.

Clinical Picture

Some reported series have found the lesion more common in males. The symptoms relate to the site of an individual area of involvement. Minimal pain and swelling prominent enough to be readily seen are features that bring the patient to the physician. When an area of the skull in involved, an area of swelling is present often with pain. The swelling is soft and not particularly tender to palpation. Unilateral exophthalmos or peri-orbital swelling may result from lesions in this area. Long bone involvement is more likely to be featured by pain.

There is little evidence of generalized illness in the patient when first seen. Loss of weight and anorexia follow more severe and multiple areas of involvement. The laboratory findings are not remarkable. Lichtenstein and Jaffe noted an eosinophilia in from four to ten per cent in their cases. There may be a mild leukocytosis. The calcium phosphorus phosphatase total protein and blood cholesterol are usually normal.

Differential Diagnosis

The cystic lesions of bone must be considered. These include solitary bone cyst giant cell tumor and benign growth defects. Solitary bone cyst is associated with the epiphyseal line appears to be derived from it and is not subject to remodelling. Giant cell tumor occurs in the early adult age group. Growth



Figure 437 Eosinophilic granuloma of the tibia revealing cystic like appearance with total destruction.

defects are usually linear rather than rounded and not associated with bone reaction.

Osteomyelitis, tuberculosis and syphilis should be borne in mind. Osteomyelitis is accompanied by the clinical and laboratory evidence of infection. When eosinophilic granuloma involves a vertebra, the differentiation from tuberculosis by roentgenogram may be quite difficult since the abscess shadow associated with tuberculosis may be duplicated by this lesion. Compere has pointed out eosinophilic granuloma as producing the picture of Calves disease in the vertebra.

Lesions such as Ewing s tumor and leukemia are ordinarily characterized by an infiltration type of destruction rather than the total destruction featuring eosinophilic granuloma. Multiple myeloma is an adult disease The differentiation from solitary bone cyst may be impossible and awaits the result of bone biopsy

Figure 438 Eosinophilic granuloma of the skull with minimal bony reaction

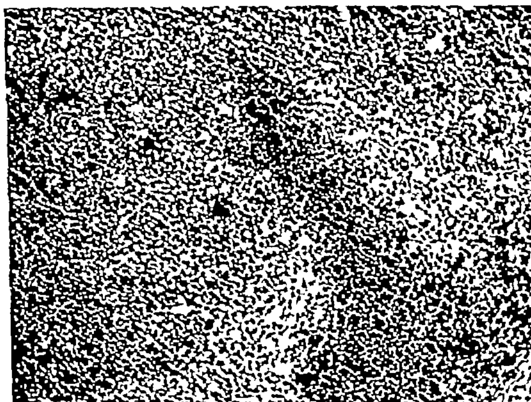


Figure 439 Eosinophilic granuloma with dark staining eosinophiles distributed ov[er a]
background of the pale staining histiocytes

Treatment

The initial or most accessible lesion is biopsied and diagnosis follows correlation of the patient's examination, laboratory data, history and the gross and microscopic findings The rapidity with which lesions can appear and also disappear with treatment is impressive

Once the diagnosis is established the choice between curettage and roentgen therapy will rest with the location of the lesion. Both methods are very efficacious in removing the individual lesions In areas where other structures, such as epiphyseal lines and ovaries may be affected by roentgen therapy, curettage is favored. Accessibility of the lesions and gravity of the procedure involved is another consideration.

The course of the patient cannot be predicted at the outset. New lesions may appear while the original lesions are being treated As long as the entity is confined to bone the prognosis is good. The ever present possibility of Hand Schüller-Christian disease exists, however, and should result in a guarded outlook.

RESISTANT RICKETS AND OTHER DISORDERS OF CALCIUM AND PHOSPHATE METABOLISM

ROBERT KLEIN M.D

Patients with resistant or refractory rickets constitute a problem which is so common that all orthopedic surgeons dealing with children should be conversant with the disease and its manifestations. Indeed in most pediatric clinics, cases of ordinary vitamin D deficiency rickets constitute only about one-half of the total number of cases of rickets Unless patients with refractory rickets are discovered early enough they become severe orthopedic problems. Six new cases of refractory rickets were found in the last five years at the Children s Hospital of Pittsburgh. During this time there were six cases of renal rickets and three cases of hepatic rickets At the same time there were twenty cases of vitamin D deficiency rickets. There is no point in applying orthopedic treatment to patients with resistant or other forms of rickets until the metabolic disease is brought under complete control. Therefore before refractory rickets is discussed as a clinical entity it is important that the metabolic changes in this and related diseases be considered No mention of the x ray changes or the appearance on histologic section of the various forms of rickets will be considered in this chapter since it is in general fair to state that neither gives sufficient evidence to determine etiology 'Rickets is rickets" when studied by these methods. In patients with vitamin D deficiency rickets rickets associated with total renal failure rickets associated with renal tubular disease and hepatic rickets therefore the metabolic and clinical changes must be understood so that the proper diagnosis can be made and adequate therapy then be carried out.

Some Metabolic Changes in Rickets

The growing infant deprived of any extra dietary source of vitamin D and kept from sufficient sunlight to manufacture his own vitamin D develops rickets

as evidenced by x ray, clinical and histologic changes His serum phosphate is low and his serum calcium is usually normal or only slightly depressed Serum phosphatase is very high. Actually his bone phosphatase is also high but this is not of clinical diagnostic importance He absorbs calcium and phosphate poorly from the intestinal tract and excretes excessive amounts of phosphate in the urine in the face of low serum phosphate values When the diagnosis is made and he is in a steady state, the magnitude of his phosphate excretion is not absolutely large. The excretion is, however relatively great in view of his low serum phosphate level His tubular reabsorption of phosphate per 100 ml of glomerular filtrate is low He has increased aminoaciduria, a decreased citric aciduria and a depressed citric acid level in his blood. When he is given vitamin D in therapeutic doses usually less than 5,000 units per day, the first response is increased tubular reabsorption of phosphate. This coupled with the increased absorption from the gut of calcium and phosphate leads to an elevated serum phosphate level. The administration of vitamin D also returns the aminoaciduria to normal and elevates the blood citric acid level and the citric acid excretion.

Action of Vitamin D and Parathyroid Hormone

It has been suggested recently that vitamin D acts in this instance by inhibiting directly or indirectly the secretion of parathyroid hormone. This theory explains most of the known facts but not all it requires ancillary postulations for whose validity there is no evidence at the present time. In any event, when extremely large doses of vitamin D are given to normal subjects or to the patient with hypoparathyroidism the vitamin seems also to have a direct renal effect. It increases the renal excretion of phosphate This action causes a lowering of serum phosphate and, coupled presumably with increased absorption from the gut leads to an elevation of the serum calcium both in patients with hypoparathyroidism and in normal persons intoxicated with vitamin D Thus, vitamin D has two different actions at different dosage levels In ordinary therapeutic doses the vitamin increases the tubular reabsorption of phosphate lessening its excretion. In high doses of the order of magnitude of 100,000 units or more per day it increases the renal excretion of phosphate In addition, the administration of vitamin D at both dosage levels leads to an increased absorption of calcium from the gut.

Parathyroid hormone may elevate serum calcium by directly causing its release from bones Vitamin D may act in a similar manner, although this has not been proven. Agents lowering serum phosphate tend to raise serum calcium. The lowering of serum phosphate lowers the product of calcium and phosphate ions and more calcium (and phosphate) are released from the bone until the point of saturation is once more reached. This is an oversimplified theory that has value chiefly as a didactic device In any event both parathyroid hormone and massive doses of vitamin D lower serum phosphate by increasing phosphate excretion and they raise serum calcium. Both these agents, in excessive amounts lead to hypercalcemia and intoxication The clinical aspects of vitamin D intoxication will be discussed later

Action of Vitamin D in Refractory Rickets

There is no evidence at present, to show whether the patient with vitamin D refractory rickets has any other defects in metabolism beyond a quantitative inability to respond normally to the usually required doses of vitamin D On the other hand there is no proof that he responds to a large dose of vitamin D in the same fashion as a normal patient responds to small doses of vitamin D The one bit of evidence against this latter possibility is the response of the patient with vitamin D refractory rickets when he is given a sufficient amount of vitamin D He heals his rickets while his serum phosphate still remains lower than normal although it does rise above the pretreatment level. This might be explained by postulating a disassociation between the two mechanisms of vitamin D action in the patient with resistant rickets. The patient with vitamin D refractory rickets may require a much larger dose of vitamin D to realize its ordinary antirachitic action At the same time, however, he may require no more than the normal subject to obtain the direct renal effect of the vitamin, which is the phosphate losing action Therefore in the large doses used to heal his rickets, the effect upon calcium absorption goes on normally, but the increased phosphate reabsorption, which would ordinarily take place is partially counteracted by the direct renal phosphate losing action of vitamin D in massive doses

Renal Phosphate Losing Rickets

Other diseases that have some of the same metabolic features as refractory rickets include the DeToni Fanconi Syndrome in which there is hyperphosphaturia, hyperaminoaciduria glycosuria and albuminuria This is basically a disease of the renal tubules and like other related syndromes there is an acidosis often accompanied by the secretion of an alkaline urine. The other renal tubular diseases are difficult to separate from each other and from the DeToni Fanconi syndrome There is a considerable argument about whether the syndrome of cystinosis is an universal concomitant of the DeToni Fanconi syndrome occurs as part of it sometimes or is an entirely separate syndrome The relationship of the DeToni Fanconi syndrome to the other renal tubular diseases producing rickets is vague. These other diseases have been characterized by Gardner as having renal base losing lesions produced either by a defect in the reabsorption of bicarbonate or in the ammonia forming mechanism. These syndromes sometimes are associated with loss of much fixed base including calcium in the urine. They are characterized by a depressed serum pH and the secretion of an alkaline urine These patients vary greatly amongst themselves in the number of symptoms they have and the combinations of these symptoms. They may show all the signs of the DeToni Fanconi syndrome including the acidosis the phosphaturia, the aminoaciduria albuminuria glycosuria polyuria and often the inability to reabsorb bicarbonate or failure to manufacture ammonia or they may show any combination of these symptoms and be classified under various eponyms.

The aminoaciduria in these diseases also tends to disappear with the administration of vitamin D and the changes in citric acid metabolism disappear Vitamin D decreases the phosphaturia, but complete healing of the rickets frequently occurs only after large amounts of fixed base have also been given to re-

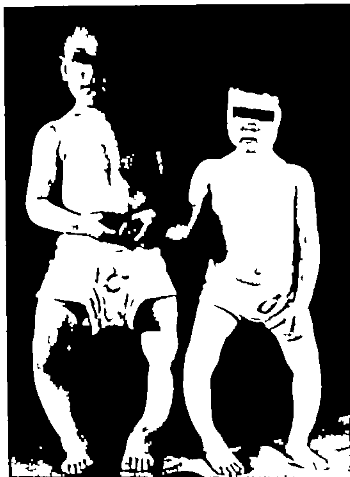


Figure 440 Siblings with bowing of the tibia in resistant rickets.



Figure 441 Rachitic changes of the distal femoral epiphyseal line This area subject to most active growth may be the only one to show changes and the last to regress.

duce the acidosis In general, the rickets in these patients is not so severe as to require orthopedic care. Perhaps this is because many of them die early with uremia It is more likely, however, that the rickets is mild because they grow very poorly and very slowly and *rickets does not become apparent frequently* in the child who does not grow However, they do suffer from osteomalacia as does the nongrowing adult who is on a diet poor in calcium and is deprived of vitamin D and sunlight over a long period of time Milkman's syndrome may occur in the later stages of these diseases but occurs more frequently in patients with the syndrome to be described next.

Renal Phosphate Retaining Rickets

Patients with a diagnosis of 'renal rickets' contrast with those whose rickets is associated with a tubular defect. This group of patients is made up of individuals with chronic renal failure of diverse etiology They present all the classic signs of chronic renal failure. They have the continuing acidosis and a retention of nitrogenous products marked by a high blood urea nitrogen or non protein nitrogen. In *addition, they have a high serum phosphate due to depressed* glomerular filtration This is frequently accompanied by a lowered serum calcium. This situation stimulates a relative or secondary hyperparathyroidism and in a certain number of these patients, the calcium is maintained at normal levels. Coincident with this they develop signs of rickets. The excess parathyroid hormone apparently causes the release of calcium and phosphate from the bones This leads to a failure to calcify osteoid and prevents the orderly change of growing cartilage to bone This is characteristic of rickets. The acidosis contributes to the leaching of calcium from bone. Furthermore there may be an excessive loss of fixed base, including calcium, in this condition which also contributes to the development of the rickets.

Patients with rickets associated with renal tubular defects such as the DeToni Fanconi syndrome frequently progress to chronic renal failure. They are then impossible to distinguish from the patients who are in the group usually characterized as "renal rickets." The nomenclature is difficult and at times is confusing. However a diagnosis of "renal rickets" usually implies a patient with rickets associated with chronic renal failure and signs of uremia in greater or *lesser degrees. On the other hand* patients with specific tubular defects have no elevation of NPN or BUN and do not exhibit the signs of renal failure until late The syndromes should perhaps be regrouped as renal phosphate losing rickets and renal phosphate retention rickets or some variant of this. Admittedly *there would be an overlap between the groups* when the phosphate losing patients developed uremia.

Hypercalcemia

Nearly all of the changes of vitamin D intoxication are duplicated by hyperparathyroidism. *The latter seems to present a higher incidence of gross renal* calculi and bone rarefaction but this may merely be a question of chronicity When large doses of vitamin D are given by error or are given for their pharmacologic action on the kidney and there is an overdose the first sign associated with the hypercalcemia is usually anorexia followed by constipation Polyuria

accompanied by polydipsia occurs very early The specific gravity of the urine is low and if the child is very small or for some reason fluid cannot be ingested, dehydration may supervene. Nausea occurs early As the toxic symptoms progress there are vomiting and signs of nervous system change accompanied by irritability, lethargy and extreme flaccidity of the muscles Apparently, there is actual relaxation of the ligaments because these children frequently have hyperextendible joints in addition to flaccidity As the disease progresses, metastatic calcification may occur Further signs of renal damage are produced, which may result from calcification of the kidney parenchyma, although presumably the earlier renal changes are merely a further manifestation of the pharmacologic or direct toxic action of the high doses of vitamin D upon the kidney The renal damage may be manifested by an elevated blood NPN but this elevation, in part, may be due to the dehydration that occurs The skin becomes sallow and pale and mottles readily on exposure to room temperature. At times it is dry and coarse Metastatic calcification occurs elsewhere throughout the body It is apparently easily found by slit lamp examination just beneath the conjunctival tissues.

Hypercalcemia may also be produced by immobilization of even the normal child or adult. The child, of course, is less likely to be immobilized unless he is actually in a cast or some other restraining device. However, even the comparative immobility of enforced bed rest will lead to a release of calcium and phosphate from the bones of the normal individual. This may at times be great enough to produce an hypercalcemia sufficiently severe as to cause symptoms Renal stones may be produced by the attendant hypercalcemia

Refractory Rickets

Refractory rickets is an inherited disease In the clinic of Children s Hospital of Pittsburgh there are several families of three generations who are being followed. The disease can be traced back in these and others for several more generations This is a common occurrence in all such clinics. There are, of course, instances where no previous manifestations of such disease can be found. If the disease is not recognized in the parent the children are often considered to have rickets of the usual variety and indeed when first seen, they are not distinguishable in any way from patients with vitamin D deficiency rickets They are then treated for this condition with the usual dose of vitamin D, no response is obtained and the rickets progresses to severe deformity

It is important to arrest the disease before deformities of the pelvis are so great that the female patients are unable to deliver except by cesarean section. In the older generations the usual story is that the mother who was affected has had several cesarean sections If the disease is not recognized osteotomies are frequently done for the correction of the deformity of the legs without the administration of other than small doses of vitamin D The osteotomies heal well and the immediate results are good but unless the rickets is controlled the deformities recur as severely as before It is imperative that the diagnosis be made early so that deformities can be prevented If deformities have already occurred the disease must be brought under control before reparative surgery is carried out.

If when the patients are first seen, they are about a year of age, there is no way to distinguish refractory from vitamin D deficiency rickets. The physical findings in both conditions are the same, as are the chemical findings of a normal or slightly low serum calcium, a low phosphate and an elevated phosphatase. One is suspicious of the refractory disease when one sees patients with rickets who are several years or more of age. Suspicion is confirmed when the patient fails to respond to the ordinary therapeutic dose of vitamin D. At this time it is necessary to consider the various types of rickets that do not respond to the usual treatment. These are genetic refractory rickets and the rickets of the various renal diseases. (Chronic hepatic disease and sprue-like syndromes can also cause rickets but the etiology should be apparent whenever the rickets occurs and will not be considered here.)

Renal function should be investigated first. Simple urinalysis is exceptionally helpful in these diseases. The patient with the DeToni Fanconi syndrome has albuminuria and glycosuria, although the latter is not constantly present and repeated specimens must be analyzed. The specific gravity in all these diseases tends to be low and somewhat fixed. Since the normal obligatory urinary solute excretion coupled with the low specific gravity characteristically produces polyuria, an accurate measurement of urinary output is helpful. Where this is not possible measurement of fluid intake is useful. At times the polyuria and polydipsia are enough to cause these patients to be diagnosed as suffering from diabetes insipidus. The excretion of alkaline or nearly neutral urine in the presence of systemic acidosis is practically pathognomonic of one of these renal tubular diseases, even if more sophisticated measurements such as estimations of the ability to form ammonia or of bicarbonate reabsorption, cannot be done. Therapeutic responsiveness is of no help in distinguishing between the various renal tubular diseases and genetic refractory rickets. Both will respond to high doses of vitamin D although the renal disease may require the additional administration of various alkalies.

When the diagnosis of genetic refractory rickets has been made the treatment should be carried out with large doses of vitamin D by mouth although it can be administered intramuscularly. Unless there is some failure or difficulty in gastrointestinal absorption such as occurs in the celiac syndrome, there is no reason why vitamin D should not be given by mouth. The required dose of vitamin D may be anywhere between 50,000 and 1 000,000 units of vitamin D per day so that the initial dose is picked arbitrarily. A dose near the lowest end of the therapeutic range should be used of course. It has been our custom to start with either 50 000 or 100 000 units a day depending upon the size of the child. If there is no response as evidenced by rising serum phosphate, falling serum phosphatase or later by roentgen evidence of healing of the rickets the dose is raised. We check the serum calcium, phosphate and phosphatase values at least once a month and also at this time do a routine urinalysis plus a Sulkowitch test on the urine. An increasing Sulkowitch response should be a warning of any hypercalcemia and is a help in avoiding serious signs of vitamin D intoxication. Healing occurs in the absence of a normal serum phosphate. Usually the serum phosphate is maintained just over 3 mg. per cent. This is higher than in the untreated patient but is lower than in the normal growing child. Apparently

this represents a balance between the pharmacologic effect of vitamin D in huge doses, that is phosphate excretion, and the more physiologic effect of small doses of vitamin D in the normal, that is the increased tubular reabsorption of phosphate. When rickets is controlled, the serum calcium value rises to a normal if it has been slightly low, and the serum phosphatase falls within the normal range. It is difficult, at times, to maintain healing of the rickets without permitting a small amount of hypercalcemia. The patient with controlled refractory rickets usually has a higher serum calcium than before treatment, frequently at the top of the normal range or just above.

The goal in the treatment of refractory rickets is to find a dose of vitamin D that is large enough to maintain healing of the rickets and at the same time one that does not produce enough hypercalcemia to cause symptoms. In our clinic the average well controlled patient with refractory rickets has a normal serum phosphatase but has a calcium that averages 11 mg. per cent and a phosphorus that averages just slightly over 3 mg. per cent. The slight hypercalcemia which he may occasionally have gives no symptoms. He eats well, gains weight and, in general, looks much healthier than before. His color is improved. Although these patients begin to grow better after they are treated, even the treated patient tends to grow less rapidly than the normal and as adults they tend to be short. However, they are not as dwarfed as the grotesque untreated patient with this disease. The excessive lordosis and potbelly of rickets disappear with treatment and, as growth continues, the visible and reparable stigmata of rickets disappear.

If treatment is carried out late, when the child is in a period of slow growth, all of the bony deformities will never disappear under vitamin therapy. These, then, are the paitents who will require osteotomy and other surgical care. One statement that must be taken with a grain of salt concerns the eventual height of these patients, since in this clinic none of them had been diagnosed before some deformity was visible and they had already lived beyond their rapid phase of growth in the first two years of life. The possibility remains that their eventual growth will be *normal if they are treated early enough.*

When they are brought in for osteotomy, the vitamin D in these patients should be stopped because immobilization will tend to give them hypercalcemia and this may produce serious kidney damage. After the osteotomy, the patient should be mobilized as soon as possible. Exercises or as much motion as possible should be carried out even when the child is in bed in a cast. Renal stones in these patients are avoidable.

After the patients have been controlled for a period of time the dosage required usually is fairly stable. Fluctuations, however, occur with changes in the rate of growth, changes in diet and perhaps with the variations in the amount of sunlight that the patient gets. Until the dosage is well stabilized, they should be seen at least once a month for at least a year. The interval may then be steadily increased until they are seen only once or twice a year, if good rapport is maintained with the family and they can be relied upon to report any changes early. After growth is stopped, the administration of vitamin D may be cut, probably entirely, without any real harm. Further bony deformities will not develop, although some degree of osteomalacia will exist. However, it has been

our habit to give a large but reduced dosage of vitamin D to our adult patients. We have no evidence of the value of this treatment.

SCURVY

ROBERT KLEIN M.D

The incidence of scurvy in pediatric practice, like that of congenital syphilis has become minimal. The very rarity of these diseases in some ways makes them more important for the pediatric orthopedic surgeon. Just as undiagnosed cases of congenital lues will be referred to him because of the pseudo-paralysis that some of these children have so the patient with scurvy may be erroneously considered to have primary bone or joint disease because of his refusal to sit or stand and because of the attending bony tenderness. When these diseases were more common they were less likely to be misdiagnosed and referred for orthopedic care. With the wide spread use of orange juice and ascorbic acid pills or drops, the few cases of scurvy seen are the result of ignorance or neglect.

The most common cause of scurvy in our experience has been the boiling of the orange juice by the mother the second most common cause has been iatrogenic. The doctor has usually mistakenly, supposed that the baby's eczema was due to the orange juice ingested. The mother was told not to give the orange juice but adequate provisions for replacement of this with ascorbic acid were *not made Breast milk ordinarily contains an adequate amount of ascorbic acid* unless the mother has been on a deficient diet herself Cow's milk contains approximately one-fourth as much vitamin C as human milk and after pasteurization the content falls even lower

The disease occurs most often in children between the ages of six months and one year Scurvy should be considered by the orthopedic surgeon when an infant in the latter half of the first year of life is referred to him because the baby refuses to stand or perhaps sit anymore, evidences tenderness in his legs or keeps his legs drawn up in a frog leg position. As the disease progresses the areas of tenderness may be swollen. These areas usually are the ends of the long bones. The specific symptoms are caused by the subperiosteal hemorrhages that these infants have much more often than hemorrhage elsewhere. Typical petechiae may be seen on the skin. Orbital hemorrhage occurs on rare occasions Very rarely is there any bleeding from the gums This is thought to be associated with the usual paucity of teeth at this age. However, the gums are said to be swollen at times and sore. We have not observed them. The symptoms of bone pain are also produced by the infractions that occur around the epiphyses. As the disease progresses the patients develop beading at the costochondral junctions which is very difficult to distinguish from that of rickets. In the best defined and well advanced cases the beading is much sharper and firmer and frequently is tender

Once the diagnosis is considered there usually is no trouble in establishing it. On close questioning the history of lack of intake of vitamin C is usually obtainable even though the mother may have considered that she was giving it. The differential diagnosis from trauma must be made and indeed everyday trauma probably is the cause of the infractions and some of the hemorrhages that the children with scurvy have. However in the patient with scurvy the



Figure 442 Massive ossifying subperiosteal hemorrhage in scurvy Halo like epiphyses are seen



Figure 443 Healing one year later of same femur as in Figure 442

our habit to give a large but reduced dosage of vitamin D to our adult patients. We have no evidence of the value of this treatment.

SCURVY

ROBERT KLEIN, M.D.

The incidence of scurvy in pediatric practice, like that of congenital syphilis has become minimal. The very rarity of these diseases in some ways makes them more important for the pediatric orthopedic surgeon. Just as undiagnosed cases of congenital lues will be referred to him because of the pseudo-paralysis that some of these children have, so the patient with scurvy may be erroneously considered to have primary bone or joint disease because of his refusal to sit or stand and because of the attending bony tenderness. When these diseases were more common they were less likely to be misdiagnosed and referred for orthopedic care. With the wide spread use of orange juice and ascorbic acid pills or drops, the few cases of scurvy seen are the result of ignorance or neglect.

The most common cause of scurvy in our experience has been the boiling of *the orange juice by the mother, the second most common cause has been* iatrogenic. The doctor has usually mistakenly supposed that the baby's eczema was due to the orange juice ingested. The mother was told not to give the orange juice but adequate provisions for replacement of this with ascorbic acid were not made. Breast milk ordinarily contains an adequate amount of ascorbic acid unless the mother has been on a deficient diet herself. Cow's milk contains approximately one-fourth as much vitamin C as human milk and after pasteurization the content falls even lower.

The disease occurs most often in children between the ages of six months and one year. Scurvy should be considered by the orthopedic surgeon when an infant *in the latter half of the first year of life is referred to him because the* baby refuses to stand or perhaps sit anymore, evidences tenderness in his legs or keeps his legs drawn up in a frog leg position. As the disease progresses the areas of tenderness may be swollen. These areas usually are the ends of the long bones. The specific symptoms are caused by the subperiosteal hemorrhages that these infants have much more often than hemorrhage elsewhere. Typical petechiae may be seen on the skin. Orbital hemorrhage occurs on rare occasions. Very rarely is there any bleeding from the gums. This is thought to be associated with the usual paucity of teeth at this age. However, the gums are said to be swollen at times and sore. We have not observed them. The symptoms of bone pain are also produced by the infractions that occur around the epiphyses. As the disease progresses the patients develop beading at the costochondral junctions which is very difficult to distinguish from that of rickets. In the best defined and well advanced cases the beading is much sharper and firmer and frequently is tender.

Once the diagnosis is considered there usually is no trouble in establishing it. On close questioning the history of lack of intake of vitamin C is usually obtainable even though the mother may have considered that she was giving it. The differential diagnosis from trauma must be made and indeed everyday trauma probably is the cause of the infractions and some of the hemorrhages that the children with scurvy have. However, in the patient with scurvy, the



Figure 442 Massive ossifying subperiosteal hemorrhage in scurvy Halo like epiphyses are seen



Figure 443 Healing one year later of same femur as in Figure 442.

trauma is much less than would produce these changes in a normal child. Moreover, the normal child who has undergone trauma usually will not have symmetrical lesions. The pseudoparalysis that some of these patients have suggests poliomyelitis. The true nature of the pseudoparalysis is usually easily ascertained. The painful swellings about the joints may suggest osteomyelitis, suppurative arthritis or even some rheumatoid process. However, these patients are much younger than one would expect for a patient with a rheumatoid disease. The symmetrical diffuse distribution of the scorbutic lesions helps rule out most of these processes. Scurvy is frequently accompanied by elevation of temperature which also might suggest infection. However, leukocytosis does not usually occur in uncomplicated scurvy. The pseudo-paralysis seen in congenital lues is usually seen in a younger age and is accompanied by other manifestations of syphilis. The distribution of the lesion in infantile cortical hyperostosis is unlike that of scurvy involving the mandible, clavicle and ulna chiefly. There is no hemorrhage and the x ray is diagnostic of this syndrome.

Finally the roentgen appearance of the bones should confirm the diagnosis of scurvy. The most dramatic changes on x ray are those in early healing when new bone begins to form beneath the elevated periosteum. The periosteum itself, is of course, invisible on x ray. At times this has suggested bony tumor. More important for clinical diagnosis is the ground glass appearance of the long bones due to the atrophy of the trabeculae. The cortex is thin and the zone of provisional calcification shows increased density beneath which there is zone of rarefaction. Infractions occur at the epiphyseal line and actual displacement of the epiphyses occasionally is seen. Calcification is seen extending out as spurs from the epiphyseal line where the periosteum is attached.

There are various tests to prove the diagnosis of ascorbic acid deficiency. Perhaps the best of these is the measurement of ascorbic acid content of the buffy coat of centrifuged blood.

Treatment of the disease is simple, requiring the administration of ascorbic acid and very rarely is there any local treatment of the extremities needed. The normal contours of the bones may not be seen by x ray for many months or even a year as the subperiosteal bone formation has been great. The subsidence of symptoms with the onset of treatment is very rapid and very dramatic.

HEMOPHILIA

The transmission of hemophilia occurs through the mother and is seen as a clinical entity in the male. It results in prolongation of the coagulation of the blood. While it may produce unsuspected surgical complications, it produces orthopedic pathology due to the frequent hemorrhages into the joints and muscle which accompany some cases. The disease does not change with age. The orthopedic complications include contractures, degeneration of the joint surfaces, pseudotumors and loss of circulation to the femoral head.

Etiology

This disease is transmitted via the X-chromosome. Since this is a sex determinant, the disease is sex linked. The gene is recessive and rare. It results in the female transmitting the disease which occurs as a clinical defect only in the male.

Clinical Picture

There is usually a past history of exaggerated bleeding in response to trauma The same patient tends to return again and again with joint hemorrhages They usually occur in the same joint The knee elbow and ankle are most frequently involved Some youngsters with hemophilia appear to escape the predilection for joint hemorrhages as a complication About one-third of the cases are repeatedly subjected to joint hemorrhages There is usually antecedent trauma although it may not be of sufficient degree to cause joint hemorrhages in a normal individual

There is joint swelling heat and tenderness at the knee with limited extension of the knee Hemorrhage into the hip results in a hip flexion contraction, and pain on attempted motion away from a position of comfort Winston has emphasized that the hip may be subject to loss of circulation of the femoral epiphysis resulting in an x ray picture resembling Legge-Perthes Disease.

Bleeding may take place into the muscles of the thigh or calf resulting in painful tense swollen and tender areas In the calf such hemorrhages into enclosed fascial spaces may threaten the vitality of the muscles and lead to Volkmann's contracture type of involvement Necrosis of muscle and loss of function may follow Ghormley has called attention to so-called pseudotumors" which may arise as further bleeding occurs into a clotted area

Repeated joint attacks lead to the development of permanent contractures. Such joints become then even more liable to further hemorrhages as the joint is used in an awkward and strained position.

Laboratory findings reveal that the coagulation of the blood is prolonged over the normal six to fourteen minute clotting time and may take several hours The bleeding time is not prolonged Platelets are normal. The hemoglobin and red cell count may be reduced. A leukocytosis may accompany the hemorrhage.

Roentgen Picture

An acute hemorrhage may reveal encroachment on the joint area of shadows of water density which fill the anatomical outline of the joint. The joint space may be widened. Further and repeated hemorrhages may eventually result in changes suggestive of degenerative arthritis of the joint with destruction of the joint surfaces The destroyed areas are often central and there may be areas of radiolucency in the subchondral and epiphyseal bone. These cavities apparently due to intra-osseous hemorrhages are frequently seen

Pathology

Repeated hemorrhages result in a condition of the synovial membrane best described as 'hemosiderosis The loose ordinarily absorbent areas of the subsynovia become clogged with hemosiderin pigment The synovia presents a bluish or purplish discoloration on examination. The joint cartilage may be separated from the subchondral bone by hemorrhage and there may be intra osseous hemorrhages as well. The articular cartilage may be thinned or destroyed with attempts at fibrous tissue replacement. Key has brought out the fact that this process tends to take place centrally

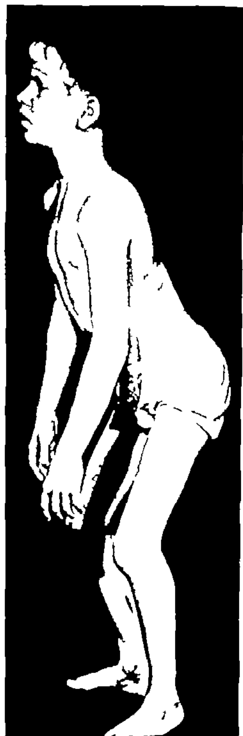


Figure 444 Knee flexion contractures in patient with hemophilia.



Figure 4.5 Roentgenogram of knee with flexion contractures in hemophilia

Treatment

The hemophiliac in the phase of acute bleeding can have the clotting time reduced in the majority of cases by the infusion of fresh blood or blood products. This includes fresh blood and plasma or the isolated Cohn s fraction I

The care of the joint containing blood in a patient with hemophilia is difficult Because of the development of hemosiderosis of the synovial membrane with repeated hemorrhages every effort should be made to reduce the volume of blood in the joint. If the clotting time cannot be reduced so that further hemorrhage will not follow aspiration there is no point in aspirating. Every effort should be made to quickly reduce the clotting time with aspiration of the joint carried out during the short period of time that the clotting time is normal. The involved limb is placed in traction to encourage motion and absorption of the blood Compression is also used following the aspiration.

The patient who has fixed joint contractures due to old hemorrhages is usually treated in traction. Such traction must be correctly applied and gently gain motion in combination with active exercises. The last few degrees of contracture may require the use of wedging casts. Such wedging must be gently and speedily done.

References

CAFFEY, J. AND SCHLESINGER, E. R. Certain effects of hemophilia on the growing skeleton. Some roentgenographic observations on overgrowth and dysgenesis of the epiphyses associated with chronic hemarthrosis. J. of Pediat., 16: 549, 1940.

COLLINS, D. H. Haemosiderosis and haemochromatosis of synovial tissues. J. Bone & Joint Surg., 33B: 436, 1951.

GHORMLEY, R. K. AND CLEGG, R. S. Bone and joint changes in hemophilia. J. Bone & Joint Surg., 30A: 589, 1948.

KEY, J. A. Hemophilic arthritis (bleeder's joints). Ann. Surg., 95: 198, 1932.

WINSTON, M. E. Haemophilic arthropathy of the hip. J. Bone & Joint Surg., 34B: 412, 1952.

GENERALIZED AFFECTIONS

FIBROUS DYSPLASIA

Fibrous dysplasia is a disease entity which may involve one or many bones. It may be combined with abnormal pigmentation of the skin, precocious sexual development, and early skeletal maturation. Still other endocrine based abnormalities may be present, and varying combinations of affects and areas of bony involvement may be present in any one case.

The disease appears to be a developmental defect. It is most common in childhood but may occur in adults; the adult lesion tends to be monostotic. It ranks next to multiple exostoses in incidence as a widespread anomaly of skeletal development. It is more common in the female.

Clinical Picture

The disease may be varied from the clinical point of view. It appears possible to divide it into three general types.

I. Involvement of a Single Bone

The lesion is usually insidious in onset unless a weight bearing bone is involved which may lead to pain and limp. Rib, clavicle, maxilla, tibia or femur have all been involved in this monostotic form which comprises approximately one-third of the reported cases. Limitation of joint motion may be present should areas such as the femoral neck be involved. Expansion of the bone may be evident on examination and localized tenderness found. A pathological fracture may first bring the patient to medical attention.

II. Lesions in Several Bones

Skin pigmentation of the cafe au lait type may or may not be present. About thirty five per cent of the ninety cases in the literature summarized by Jaffe and Lichtenstein showed a definite degree of cutaneous pigmentation.

III. Multiple Lesions

Pigmentation of the skin and precocious puberty—these cases represent the most severe form of the disease. Cases in which the lesions tend to remain unilateral may be regarded as less severe than the completely widespread type. The more severe forms are found in younger children and infants, and involvement of the skull is common. This form of disease may lead to serious deformities and crippling with repeated pathological fractures in the past history.

Only the female appears to be subject to precocious puberty with this disease. Rarely hyperthyroidism has also been found accompanying the bone lesions. The laboratory findings have usually been within normal limits. An occasional exception is the alkaline phosphatase which is elevated beyond that expected for the age.

Roentgen Findings

Areas of decreased density appear in the bone. There is a ground glass appearance of the involved area which decreases the contrast with the surrounding bone and aids in distinguishing the lesion from a single bone cyst. The lesion spares the epiphyses.

With growth the cortex may become attenuated and expanded. As involvement becomes more generalized the entire long bone reveals the thinned cortex and fine trabeculae somewhat reminiscent of osteogenesis imperfecta. There may be bone deformity secondary to previous fractures. Generalized involvement of the bone may become the most striking feature rather than localized cyst like areas. Rapid extension of the condition is not characteristic. Indeed many localized lesions have been observed dormant for years.

Sclerosis of any marked degree about the lesions is not characteristic. In the generalized disease the base of the skull may appear sclerotic, however. There is in addition replacement of normal contrasting bone with areas of "ground glass appearance in varying portions of the remainder of the skull.

Pathology

Lichtenstein and Jaffe have been responsible for clarifying the pathology of this condition under the heading fibrous dysplasia. When a localized lesion is approached surgically the cortex overlying it is frequently found to be eroded from the medullary side and more vascular than normal.

The contents of the area consist of a firm compressible tissue generally whitish with occasional red specks. The tissue is primarily connective tissue perhaps gritty to cut if bone formation is prevalent within it and occasionally containing areas of cartilage. This tissue may form virtually the entire composition of the medullary portion of the bone. Rarely some cystic softening of this tissue may be encountered.

Microscopically such tissue is composed of small spindle shaped cells in a loose and whorled arrangement. The cellularity varies considerably. Scattered irregularity in this tissue may be primitive osseous elements formed by metaplasia of the connective tissue. This osseous tissue is atypically calcified or frank osteoid. The tissue tends to be avascular except in highly cellular areas. Small

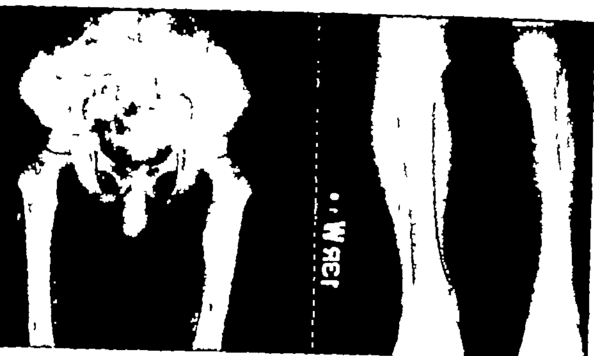


Figure 446 Fibrous dysplasia of femur and tibia

Figure 447 Replacement of medullary cavity contents in bone by proliferating fibrous tissue and osteoid in fibrous dysplasia.

islands of hyaline cartilage may also be found The presence of the cartilage foci aid in the certainty of the diagnosis

Treatment

Although the disease particularly in severe forms, tends to begin in infancy and childhood it is the general rule for the patient to survive into adulthood A surgical attack in an isolated lesion with curettage and replacement of the area with bone chips is usually followed by eradication of the area In the more florid and widespread form of the disease, surgery is reserved for those lesions which by location or extent are a source of clinical trouble. In widespread areas pointed out by Strassburger, Garber and Hallock the repair process may fall before the spread of the lesion and the bone chips be resorbed

Isolated lesions may be attacked for diagnosis, but it should be remembered that with cessation of growth the lesion appears to diminish in activity and become dormant

Reference

LICHTENSTEIN W AND JAFFE, H Fibrous dysplasia of bone Arch. Path., 33: 777

OSTEOPETROSIS

This condition was described by Albers-Schönberg in 1904. It is characterized by a homogeneous opacity of the bones with obliteration of the marrow cavities and a tendency to loss of trabecular structure The bones may be abnormally fragile An accompanying myelophthisic anemia is the rule

The disease is rare and has been found in both male and female There is a strong familial tendency It has been found in the fetus and in childhood and in adults The cause is unknown

The disease in childhood is progressive with anemia hemorrhage and infection usually leading to an early fatal outcome Some cases do run a more benign course and on reaching adulthood the disease tends to remain stationary According to Nussey the condition is benign when inherited and malignant when there is consanguinity of the parents The children tend to be retarded in all phases of development with a degree of dwarfism usually present In addition to the anemia optic atrophy deafness and facial or ocular palsy is often found. The blood chemistries do not show any characteristic changes although the serum calcium is occasionally raised. Analyses of the bones indicates an increased calcium and phosphorus content.

Cohen reviewed the pathology in this disease He found all bones of the skeleton affected Membranous bones showed changes analogous to cartilaginous ones Cartilage cell columns continued to mature but there was a disturbance of the normal sequences of resorption of the primary spongiosa Thus there appeared to be a great quantity of unresorbed cartilage matrix upon which the usual process of deposition of bone and osteoid went on without the matrix being resorbed

The rate of fracture healing is unusually rapid but there is marked delay in the remodelling sequences associated with growth There is no tubular marrow

Figure 448 Osteopetrosis with pathological fracture of the femoral neck

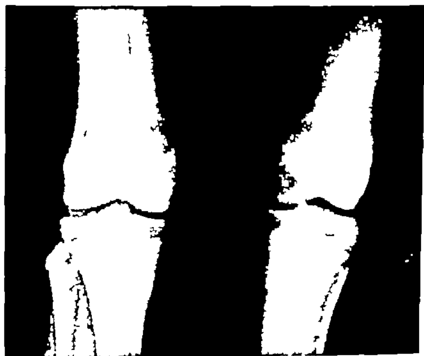


Figure 449 Osteopetrosis with hindrance to remodelling of the metaphyseal areas of the femur and tibia.

cavity The cartilage and bone matrix appears well calcified, but there are large areas of osteoid formation. Cohen noted that the transverse lines in the metaphyses of osteopetrosic bone were fractures in various stages of healing.

By x ray the bones appear to have an increased opacity and loss of structure within their outline The metaphyses show a failure of remodelling so that they are clubbed or box like Transverse dense lines are frequently apparent and occasionally separate metaphysis from diaphysis. The skull shows increased density most marked at the base.

The bones appear solid on cross section and grey white in color No fatty marrow is seen The trabeculae are increased in number and thickness Some nodules of cartilage may persist among the trabeculae As Cohen suggests the delay in remodelling results in the formation of chondro-osseous complexes, absence of a marrow cavity and may also account for the clubbed metaphyses

The treatment revolves around aid for the ever present anemia which may vary in severity with exacerbations and remission The child with unduly fragile bones is frequently seen with fractures. The duration of immobilization should be shortened compared to the normal No cure of the fundamental condition is known

References

Cohen J Osteopetrosis case report, autopsy findings and pathological interpretation. Failure of treatment with vitamin A J Bone & Joint Surg 33A: 923 1951

Kramer, B Yuska, H and Steiner M Marble bones II. Chemical analysis Albers-Schoenberg disease (marble bones) report of a case and morphologic study Am J Path 23: 755 1947

VanCreveld S and Heybrock, N On Albers-Schoenberg disease (marble bones) Acta Paediat., 27: 462 1939

OSTEOGENESIS IMPERFECTA

This rare disease occurs in a form which is present at birth and is pre-natal with the possibility of fractures and deformities developing *in utero* This form *tends to be* most severe A second form develops in post natal life, which is less severe but is characterized also by mechanical weakness of the bones and blue sclerae The disease affects both sexes but is slightly more common in the female

While the outlook for survival is poor in the hereditary pre-natal case nonetheless an individual case may survive many months, even though very severe. Milder cases may survive into adult life A tendency to improvement is frequently seen The post natal cases may have only an occasional fracture and a full life expectancy although severe cases are seen in this group also

The etiology is unknown

Clinical Picture

Another name for this disease fragilatas ossium, gives a clue to its dominant clinical characteristic It is the fractures of the skeleton that bring the case to medical attention. The relaxation of the joint capsules and the blue sclerae that frequently accompany the condition are noted secondarily The musculature is frequently loose elastic and diminished in volume.

Otosclerosis may occur in those surviving into adult life and according to



Figure 450 Involvement of the spine in osteopetrosis

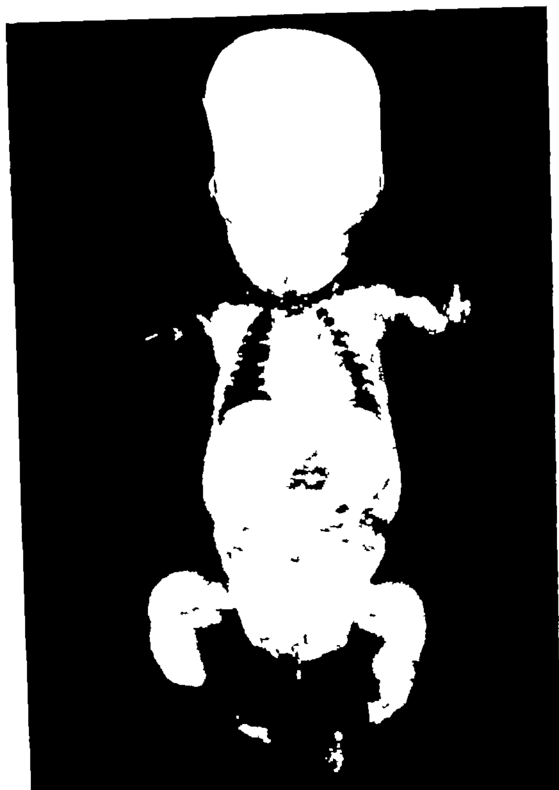


Figure 451 Infant with multiple fractures and deformity in osteogenesis imperfecta

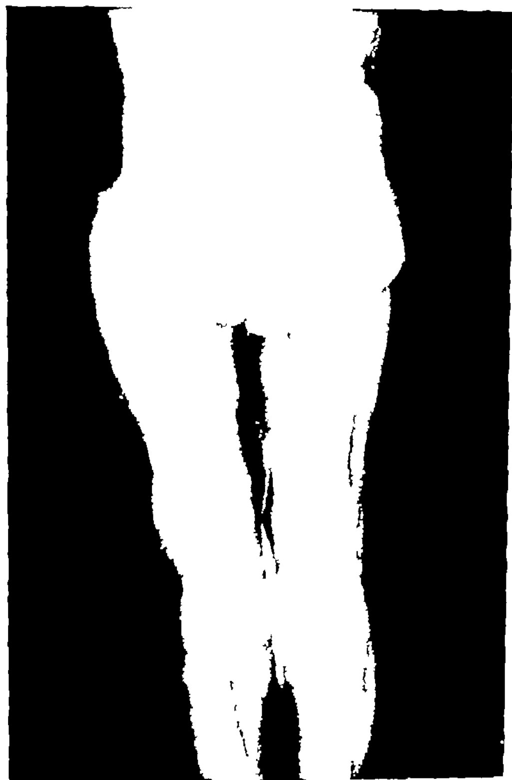


Figure 452 Older child with thin cortices in fragilitas ossium and with fracture of femoral diaphysis

Bickel *et al* is most common in the hereditary type Otosclerosis has also been found in adults with blue sclerotis without evidence of bone fragility

The more severe cases exhibit diminished growth to the point of dwarfism. The skull is unusually broad with prominent occipital and parietal bosses The teeth in childhood are poorly calcified and translucent. Blood chemistry values have not been found to be diagnostic.

Pathology

The most striking finding on bone section is the sparse distribution and the thin frail nature of the bone trabeculae The cortex is poorly formed The trabeculae are frequently osteoid. Islands of cartilage are seen and the medulla is in part fibrous or fat containing. The number of osteoblasts seen is frequently deficient.

Roentgen Findings

The most common type of bone seen is thinned with diminished trabeculation and slender cortices Varying degrees and types of deformity may be seen dependent on the past history Many transverse lines are present in the metaphysis Ossification of the skull is imperfect and patchy The fibula may be a mere thin line The vertebrae may show the effects of pressure of the intervening vertebral discs being compressed and biconcave.

Fairbank has differentiated a type in which the major long bones are short and widened but still with excessively thin cortices. Fractures are frequently seen along with deformity

Fracture Healing

The fractures in general tend to heal although non union and delayed union has been seen. Some cases tend to put out a very hyperplastic callus containing a "chondroid tissue.'

Treatment

The physician must tread a delicate line in treating fractures in this disease. Prolonged immobilization leads to further bone atrophy and the resultant hazard of further fracture in convalescence. Insufficient immobilization leads to bowing which is permitted by the soft callus Since generally muscle tone is poor, the children relatively inactive and light in weight the time of immobilization can be shortened consistent with maintaining alignment. Healing in deformity almost inevitably results in refracture

Medical measures undertaken with a view to providing a stronger better calcified skeletal system have proven of little avail

References

BICKEL, W H CHORMLEY R K AND CAMP J D Osteogenesis imperfecta Radiology 40: 145 1943

FAIRBANK, H A T Proceedings of the royal society of medicine 23: 1263 1930

FAIRBANK H A T Hyperplastic callus formation with or without evidence of fracture in osteogenesis imperfecta. Brit. J Surg., 36: 1-16 1948



Figure 453 Compression of vertebrae in osteogenesis imperfecta



Figure 464 Skull with wormian bone pattern visible in fragilitas ossium

FLEMING, B L, RADASCH H E AND WILLIAMS T Osteogenesis imperfecta J Bone & Joint Surg., 19: 725 1037
PELNER AND COHEN J N Osteogenesis imperfecta tarda. Am. J Roentgenol 61: 690 1949
WYATT T C AND McEACHEN T H Congenital bone dysplasia (osteogenesis imperfecta) associated with lesions of the parathyroid glands. Am. J Dis. Child 43: 403 1932
WRIGHT P B GLENSTETTER, S L AND GREENBLATT R. B Therapeutic acceleration of bone age in osteogenesis imperfecta. J Bone & Joint Surg., 33A: 939 1951

CONGENITAL AFFECTIONS

CONGENITAL SHORT FEMUR

There are many variations of congenital short femur in which the net result is shortening of the femur The severity varies from a mild discrepancy in which one leg approximates the length of the other to a limb in which only a rudimentary portion of the femur remains resulting in the knee approximating the inguinal fold The calf is usually surprisingly well formed although the

tibia may be somewhat shorter than the normal side. A bilateral case is not known to the author

Heredity appears to play no part in the development of the anomaly Cases are not infrequent in a children's orthopedic clinic.

Clinical Picture

The deformity is noted at birth. Examination reveals the leg length discrepancy in which the lower leg plays only a minor part. There is a great tendency for the patella to be rudimentary, and it is often dislocated laterally

Coxa vara deformity may be associated resulting in the trochanter riding above Nelaton's line A hip flexion deformity is frequently found. Muscular function is ordinarily sufficient to stabilize the joint at hip and knee.

The picture in the female is distressing There is a refusal to wear skirts or any form of clothing that will reveal the deformity The male can more readily cover the deformity and engage in normal activities Unfortunately the lesion appears to be more common in the female.

Treatment

The severity of the defect justifies radical measures particularly in the female. Those cases where the ankle of the short leg is within a sufficiently short distance of the other may be quite simply solved by appropriate epiphyseal arrests at the proper time.

Where the ankle will apparently end up closer to the knee, the underlying approach may be to secure an ankle at the level of the sound knee

This will often involve epiphyseal arrest of the short leg and fusion of the knee to stabilize it beneath the hip The foot may be trained down into equinus. The limb may be rotated partially at the time of knee fusion and later the rotation completed through the femur to gain a total of one hundred and eighty degrees (180) The calf muscles can then act as the motor for a knee prosthesis. Such a procedure was originally suggested by Barggreve (1930) Van Nes reported very satisfactory end results with this combination of events

Still other details of the deformity may need treatment. These include the coxa vara deformity and hip flexion contracture which may be associated and for which subtrochanteric osteotomy may be desirable

Reference

VAN NES C P Rotation-plasty for congenital defects of the femur J Bone & Joint Surg 32B: 12, 1950

ABSENT TIBIA OR FIBULA

The anomaly of a complete or nearly complete absence of either tibia or fibula can occur Of these absence of the fibula is most common. In addition there may be anterior angulation of the remaining bone with dimpling of the skin at the apex in the case of absent fibula. The foot is in equinus and tends to sublux laterally in the case of absent fibula or medially in the case of absent tibia. The tarsus itself may be the seat of anomalies with absence of lateral or medial rays and fusion of the tarsal bones in varying combinations. Shortening of the extremity is frequently marked.

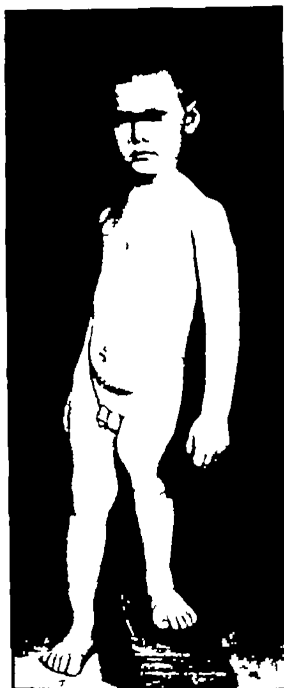


Figure 455 Congenital short femur

Etiology

The cause of the defect is unknown The embryologic time of their development is apparently the sixth or seventh week of life. Middleton felt that failure of maturation of muscle fiber resulted in failure of longitudinal growth and abnormal stress due to shortening of the posterior and lateral muscles with this stress resulting in anterior bowing and equino valgus position of the foot in the case of the absent fibula.

Coventry and Johnson in reviewing their cases of absent fibula found that heredity played a minor role.



Figure 456 The short femur and relatively unaffected tibia and fibula are seen The unossified portion of the proximal femur is at the base of the neck on the involved side



Figure 45 Congenital short femur and developmental coxa vara

Clinical Picture

The absent fibula reveals a lower leg which is short and bowed anteriorly with dimpling at the apex of deformity The foot is in an equino valgus position The absent tibia has little anterior bowing and no true knee joint with the fibula subluxated laterally There is medial displacement of the foot at the ankle. Shortening is usually present but the lack of increasing width of the proximal portion of the remaining bone aids in distinguishing the fact that it is an absent tibia rather than an absent fibular deformity

Congenital pseudarthrosis of the tibia has a similar appearance to the anterior bowing with dimpling, but the foot is not in a fixed equino valgus position

Treatment

Absent Fibula

Immediately after birth the foot is started in corrective casts to bring it into normal position beneath the tibia. This may require sectioning or lengthening of tight posterior and lateral structures in addition to conservative measures. The fibula may be represented by a fibrous cord which can be removed the peroneal muscles and tendon achilles lengthening and a capsulotomy performed at the ankle Later, once these tight structures are relieved the anterior bowing of the tibia may be corrected by osteotomy A delayed osteotomy done in two stages may be safer than a primary osteotomy although Coventry and Johnson reported no difficulty with non union in their cases. There is a problem in maintaining the foot in position. A laced foot and calf boot re-enforced with a steel brace aids in preventing recurrent deformity The shortening becomes a problem for appropriate epiphyseal arrests lengthenings or shortenings dependent on the individual case

Absent Tibia

This difficult treatment situation varies greatly from case to case. When a remnant of the tibia exists proximally the fibula may be united to it to gain a knee joint Distally the astragalus may be fused to the fibula or remnant of tibia.

References

COVENTRY M B AND JOHNSON E. W Congenital absence of the fibula. J Bone & Joint Surg., 34A: 941 1952

HARMON P H AND FAHEY J J The syndrome of congenital absence of the fibula—report of three cases with special reference to pathogenesis and treatment. Surg Gynec. & Obst 64: 876 1937

CONGENITAL AMPUTATIONS AND CONSTRICTING BANDS

So called congenital amputations exist as a single anomaly The part involved may be one digit or the major portion of an extremity It has been felt that these amputations may be a further extension of constricting bands which may be visualized as eventually disturbing the vascular system of a part *Streeter feels that they are primary germ plasm defects.*

Constricting bands which have been noted most commonly in the lower leg

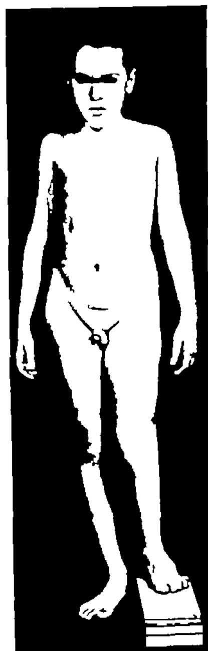


Figure 458 Absent fibula with associated shortening.

may exhibit varying degrees of severity The usual clinical picture has resulted in a tight fibrous band about the bone through which—in an incredibly small area—pass nerves blood vessels and tendons. The distal part is often enlarged with a woody edema Intrauterine amputations may still have the necrotic portion attached as in the accompanying figure.

Treatment

The treatment of the amputation consists in preventing deformity by active and passive exercise and by splinting. Such measures vary with the part involved Bracing is usually accomplished with an appropriate socket and double upright jointed prosthesis rather than an atomical appearing prosthesis until growth is through

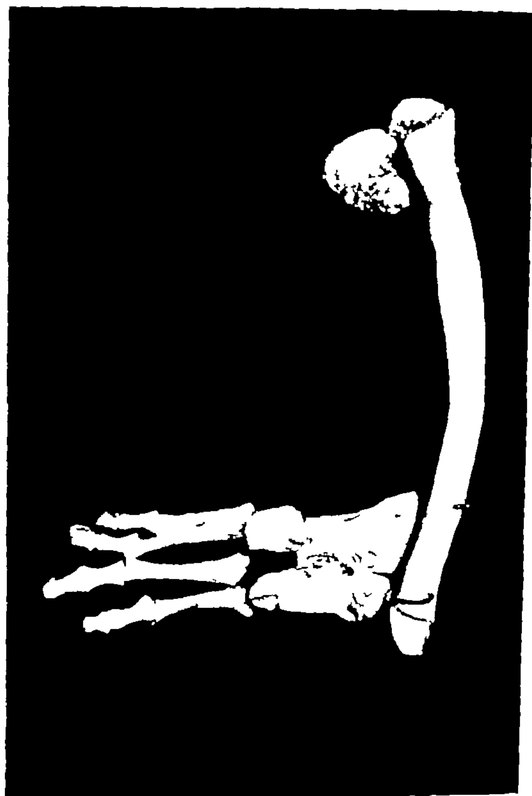


Figure 459 Absent tibia. There is a remnant of tibia remaining proximally and associated anomalies of the tarsal bones.



Figure 400 Congenital amputation The necrotic tibia and fibula are still attached to the thigh stump

The constricting band can be very satisfactorily handled if it is not all handled at once Approximately a third of the circumference of the part is done at one sitting. The incision is made on the external surface where the skin folds initially come together rather than deep in the crevice This will result in the scar remaining at skin level and not indenting Dissection must be meticulous as there is no room to spare Some superficial adipose tissue in the distal part may also have to be excised at a later date.

References

Streeter, G L Focal deficiencies in fetal tissues and their relation to intra-uterine amputations. Contributions to Embryology 126 Publication 414 Carnegie Institute Washington, 1930

CLEIDO-CRANIAL DYSOSTOSIS

Cleido-cranial dysostosis is a hereditary defect in skeletal development. It is featured by absence of the clavicles or aplasia of these bones to a degree that they are rudimentary Associated defects include an increase in the transverse diameter of the skull due to delayed union of the sutures delayed dentition of the teeth delayed ossification of the pubis deformities of the metacarpals and metatarsals and absence of bones other than clavicle.

The clavicles may be represented by a fibrous cord The patients may be short due to a decrease in longitudinal growth of long bones The teeth may not only be delayed in development but loss of deciduous teeth may be delayed. Other skeletal variations may be noted such as genu valgum, coxa vara and scoliosis

Clinical Picture

The usual clinical picture seen is involvement of the clavicles and bones of the skull. The sutures are widened at birth the fontanels gaping and the clavicles absent. The picture may vary and various combinations of defects be present. The shoulders can be brought together anteriorly without the clavicles. The upper portion of the thorax is narrowed and flattened

The patient is symptomless. Motor function for the most part is not interfered with and the patient is not handicapped.

Treatment

Treatment is ordinarily not indicated. Should deformity produce disability, it is corrected on an individual basis.

8

Affections of Muscle

PSEUDOHYPERTROPHIC MUSCULAR DYSTROPHY

Pseudohypertrophic muscular dystrophy, a progressive condition of loss of muscle function is seen quite commonly in orthopedic clinics. It is a primary myopathy The cases are often hidden among those reporting because of flat feet.

The disease is frequently familial and the frequent incidence in male siblings is familiar The onset usually takes place before the age of six. The weakness is symmetrical progressive and featured by enlargement of muscle groups of which the most striking is those of the calf At the same time, or slightly later, atrophy about the shoulder girdle may be noted.

Etiology

The possibility of an underlying involvement of the central nervous system has not been substantiated. A familial history can usually be obtained. The muscle defect causing the atrophy is unknown but conceded to be hereditary

Clinical Picture

The onset is insidious with symptoms first noted in the three to seven age group It is usually not appreciated by the family that the patient is weak. Difficulty in climbing stairs easy fatigue or flattening of the arch of the foot may be a complaint.

The pseudohypertrophic type is more common in males The feet are pronated and contracture of the gastrocnemius is noted early in the development of the disease. Difficulty in the gait is noted with stumbling and frequent falls. The calves are enlarged and have a doughy feel on palpation, not the hard feel of normal muscle. The child placed on the floor arises by aiding extension of the knee by the quadriceps with the hand. He then extends the hips by pushing further on the thigh.

Atrophy of the shoulder girdle particularly the pectorals may be noted on palpation. Muscle test reveals diminished muscle power particularly in the quadriceps and proximal limb muscles until late in the disease.

The fascio-scapulo-humeral type of Landouzy Dejerine is noted by early



Figure 461 Enlargement of the calves in pseudohypertrophic muscular dystrophy

involvement of the muscles of the face giving a mask like dull expression. Expression remains about the eyes longer than elsewhere in the face although difficulty is experienced in closing the eyes The lower lip hangs down and out. Atrophy of the shoulder girdle and arms is marked. Later there is involvement of the pelvic girdle

In the juvenile scapulo-humeral form of Erb atrophy of the shoulder girdle is the first form of involvement. The onset is later than the early childhood, beginning in pseudohypertrophic muscular dystrophy and usually occurs in the teen age group The reflexes are absent in those muscles severely involved.

Late forms of muscular dystrophy with pelvic girdle atrophy show a gluteus maximus and medius limp. Scoliosis is found with trunk weakness and is progressive even in those patients confined to a wheel chair

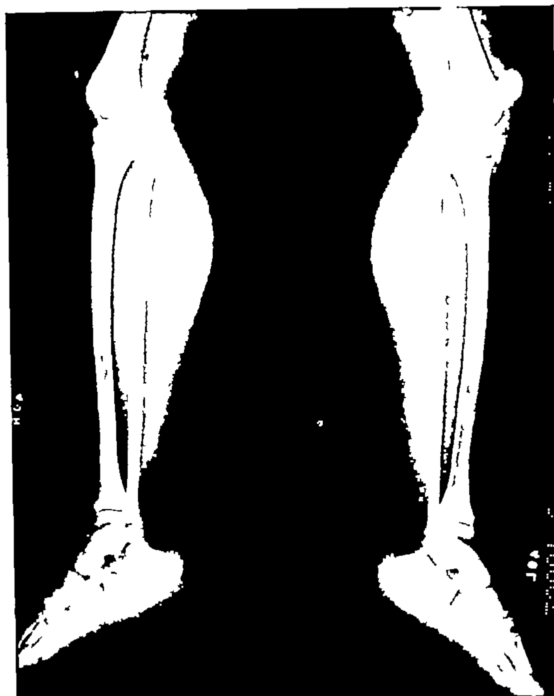


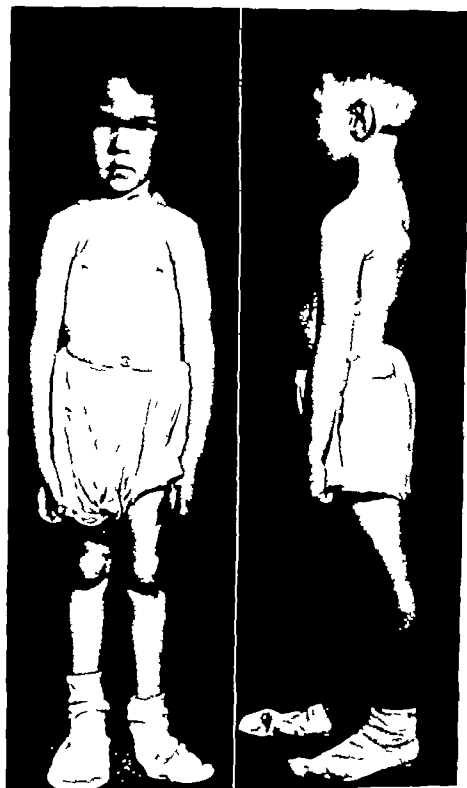
Figure 462 Roentgenogram of the calf enlargement with streaks of fat density Enlargement is limited initially to this area.

Pathology

The essential muscle changes are similar in all types of dystrophy The gastrocnemius in a marked case of hypertrophy looks fatty The muscle color varies from a grayish and pale to yellowish.

There is considerable variation in fiber size fatty replacement and connective tissue invasion of the muscle Swelling of the fiber is apparently the first change

Sarcolemmal nuclei are increased in number and may be increased in size. In



Figures 463 464 Scapulo-humeral form of muscular dystrophy

some forms of dystrophy the nuclei may occupy a central position in the fiber Denny Brown has noted that the biopsy site if rebiopsied ten to fourteen days later exhibits degenerative changes with loss of myofibrils without evidence of regeneration of muscle buds In later cases vascular and granular degeneration of the fibers is found. Changes in sensory and motor nerve fibers are not seen and changes in late disease are relatively minor and do not point toward a neural atrophy

Treatment

A specific treatment to stop the onward march of the disease has not been found. Various medications have been felt to be helpful at least for periods in the patients' course. These include glycine by mouth and subcutaneous injections of epinephrine and pilocarpine. Alpha tocopheral (Vitamin E) has been found to inhibit the creatinuria in some of these patients. A program of resistive exercises for key muscles such as gastrocnemius, quadriceps and gluteus maximus has been helpful providing the course is periodically interrupted to allow the patient to freshen his interest.

Stretching exercises are particularly necessary for the gastrocnemius. The tendency for contracture is marked providing an additional handicap in the already overburdened patient. Tenotomy of the tendo-achilles is rarely necessary to release such a contracture. Bivalved night casts in the corrected position may be an aid in prevention.

Prolonged periods of bed rest must be avoided as further weakening the patient. Muscle biopsy of the gastrocnemius when done to confirm the diagnosis, should be followed with a protective splint and early weight bearing to prevent contracture.

As the trunk begins to sag back braces are frequently necessary, the Barr Bushenfeldt type with lateral arms being particularly helpful in preventing lateral slump in a generally weakened trunk The Milwaukee type with chin and occiput support may also be helpful

References

ADAMS R. D DENNY BROWN D AND PEARSON C M Diseases of Muscles. Paul B Hoeber Inc New York, 1953

DENNY BROWN D The nature of muscular diseases. Canad. M A J 67: 1 1952

LANDOUZY L. AND DEJERINE, J De la Myopathie atrophique progressive Myopathie sans neuropathie debutant d ordinaire dans l enface par la face Rev de Med 5: 81 253 1885

ERB W H Über die "juvenile Form" der progressiven Muskelatrophie ihre Beziehungen zur Sogenannten Pseudohypertrophie der Muskeln Deutch Arch Klin. Med 34: 467 1884

DERMATOMYOSITIS

This is a rare highly disabling disease whose activity leads to marked contractures and with involvement of muscles of respiration and deglutition may lead to a fatal outcome. The activity of the disease is marked by a non suppurative inflammation of striated muscles usually accompanied by skin lesions.

Etiology

Attempts have been made to implicate parasitic viral and bacterial agents but these have not been substantiated. No definite nutritional disturbance has been found. The disease has been grouped with so-called "collagen diseases" such as scleroderma.

Clinical Picture

The onset is usually insidious with weakness and chronic fatigue. The development of limb stiffness and muscle pain follows these initial symptoms. Some cases go on to develop considerable weakness which is usually irreversible if extensive. The muscle soreness leads to flexion contractures of the joints.

A low grade fever is present at some time in the course of the disease. Skin involvement of the face may be present early in the disease. The extremities tend to become wasted with a brawny inelastic feel Shiny or scaly skin tends to be drawn tightly over bones and atrophied muscles. The atrophy is extensive

A fatal termination if that is the course usually takes place in the first two years the disease thereafter tending to become quiescent.

Pathology

A biopsy of skin, subcutaneous tissue and muscle taken in continuity from one area may be helpful in establishing the diagnosis. Early changes show edema of subepithelial tissues and collections of polymorphonuclear leukocytes in both skin and muscles. These leukocytes are mainly eosinophils. Later foci of lymphocytes and monocytes are seen, particularly at the junction of fascia and muscle. Muscle necrosis may be seen In the late stages atrophy and disappearance of muscle fibers without inflammation is usual.

Treatment

The measures to combat the disease revolve around the prevention of deformity and the compensation for muscle weakness Periods of rest in the corrective position and bivalved casts, if the tendency to deformity is great, are essential. Use of casts must be balanced by periods of exercise out of casts during the day For existing deformity traction splints corrective operations and stretching by active and passive manipulation may all be necessary

Improvement is characterized by increase in muscle strength, decrease in fever if present and a subsidence of skin lesions and muscle soreness and with a gain in body weight. Active exercises of a resistive type may be given when the muscles are no longer sore.

Medication has ranged from hormones including adrenocorticotropic hormone to Vitamin E Results have not been encouraging but occasionally suggestive improvement has occurred which is difficult to evaluate because of the tendency for spontaneous remission

ARTHROGRYPOSIS MULTIPLEX CONGENITA

Arthrogryposis multiplex congenita also known as amyoplasia congenita was first described by Otto in 1841 Its effect is to produce multiple congenital contractures in the extremities. The trunk appears to be unaffected.

Club feet, club hands and dislocated hips are common forms of involvement. The joints are fixed with extremely limited motion The patients are of normal intelligence.

Etiology

The skeletal muscles have not developed It may be that the primitive myo blast has failed to mature as suggested by Middleton. The possibility that it actually represents a neural atrophy with fatty infiltration due to primary an terior horn cell degeneration has also been suggested

Clinical Picture

The extremities appear of normal length and thickness, but on palpation have a soft feel suggestive of atrophy The full blown picture which includes equino-varus feet, club hands, dislocated hips and fixed knees may be varied by relatively isolated involvement The knees only may exhibit a fixed contracture with motion impossible in either flexion or extension Infants are seen with unilateral and isolated joint stiffness and evidence of poor muscular development about the joint which may fall into this category Such a lesion is suspected in an equino-varus foot deformity with a small calf at birth

Although any position of contracture is possible the leaning is toward ex tension at the joint Some motion is possible but usually through a very small

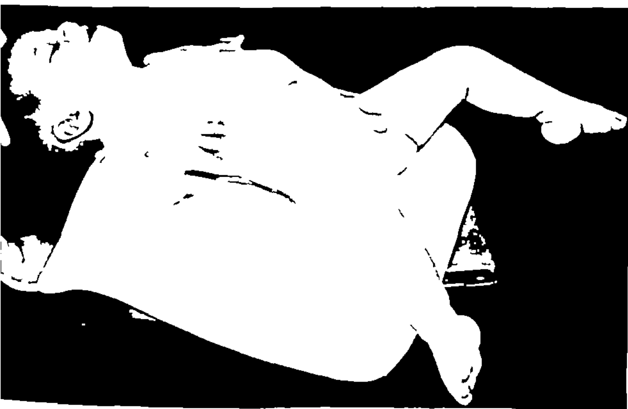


Figure 465 Contractures of hands, feet and knees in amyoplasia congenita

range. The limbs tend to be conical in form rather than developed. The arms usually are rotated inward, elbows extended and the hands flexed and forearm pronated. The hips are flexed, knees tend to be extended and the foot in an equino-varus position.

Roentgen Picture

Roentgen studies reveal the lack of normal muscle shadows in the involved extremity with thin strings of water density running through the extremity which appears to consist largely of shadows of fat density

Pathology

Adams, Denny Brown and Pearson in their text point out that in one of their cases of hip flexion contracture the flexors of the hips were well developed, but the extensors atrophied, lending some credence to a neurological etiology Some muscles can not be found at all in masses of fatty tissue others are represented by occasional rudimentary pale, pink fibers. The small size of the muscle fibers seen is noteworthy Degeneration of muscle fibers is not a marked finding.

The joints themselves are unaffected with normal appearing cartilaginous surfaces It is apparent that the cause of limited motion and contracture resides in the soft tissues In the cord the anterior horn cells are markedly reduced in number and some areas may be completely absent.

Treatment

The treatment consists of whatever means are necessary to correct deformity and gain motion. The outlook for correcting deformity is good but improvement in motion in the extremity will be slight.

The equino-varus foot deformity is particularly resistant to correction and some times requires operative division of tight fibrous bands to gain normal alignment. The hips may sometimes prove resistant to closed reduction and operative reduction becomes necessary to secure correct alignment.

A well planned exercise program is necessary to achieve the small gains that are possible.

References

BRANDT S A case of arthrogryposis multiplex congenita. Acta Paediat., 34: 365 1947

EALING, M L Amyoplasia congenita causing malpresentation of the fetus. J & Obs. Gynaec. Brit. Emp 51: 144-146 1944

GILMOUR, J R. Amyoplasia congenita. J Path. & Bact., 58: 675 1946.

MIDDLETON D S. Studies on prenatal lesions of striated muscle as a cause of congenital deformity Edinburgh M. J 41: 401 1934.



Figure 406 Roentgenogram revealing thin strands representing muscular tissue running through shadows of fat density in the leg in case of arthrogryposis.

9

Neurogenic Affections

CEREBRAL PALSY

Cerebral palsy constitutes a major classification into which a large number of patients who can be benefited by orthopedic measures fall Unfortunately the classification of the various types is not clear The orthopedist tends to classify the patient according to his most predominant handicap From this type of classification, treatment may be more readily inferred. The neurologist tends to classify according to the probable anatomic site of the deficiency Our knowledge regarding sites of involvement in various types of cerebral palsy is still very slight.

Distraught parents seeking help for their child are driven from one physician to another until some one offers help It becomes a very difficult social and psychological problem and demands the highest standards of judgment and character from the physician. The parent and child led down a path of promised recovery only to find the child unable to compete when school or the teen ages are reached becoming bitterly disillusioned with serious consequences is not infrequent.

We must avoid castigating the physician as not being interested in the child because he insists on being on realistic ground. Too firm and definite a prognostication particularly in regard to intelligence when a child is very young is also to be avoided. Many things become evident of their own accord with growth in a slower and kinder way with much time for adjustment.

Definition

This condition covers all states in which the motor system control is interfered with by a central nervous system lesion. The principal characteristic is incoordination in the use of one or more of the extremities The side features which in some cases may become the principal feature include mental deficiency, lack of balance, convulsions and speech difficulties. The underlying etiology appears to be damage or defect of the upper motor neurone either in cord or brain.

Incidence

According to Phelps the incidence is seven cases per one hundred thousand births. Of these one dies below the age of six usually in infancy One-third of the

six remaining are mentally deficient to a degree that custodial care will be necessary One is severely handicapped, two moderately handicapped and capable of improvement and one, finally, whose handicap is so mild that treatment will not be a prominent feature With the possible exception of polio and dependent on its epidemic incidence, cerebral palsy is the most common of the treatable neuromuscular conditions

Etiology

This condition in one of its various forms stems from one or more of multiple possible etiologies

In prenatal life the brain is subject to congenital defective development and to variations in structure with resulting abnormality of function Pathological conditions present in the mother may affect the development of the fetus although cases in this category are few in number

At birth trauma may result in cerebral hemorrhage and anoxia It is very hard to separate the effects of asphyxia from hemorrhage. The frequent relation to prematurity is well known This is apparently on the basis of tissues not fully enough matured to withstand the sudden change in pressure relations at birth. Undue traction on the neck forceps pressure and birth complications such as a cord around the neck all may result in cerebral damage. Blood abnormalities may result in erythroblastosis prolonged jaundice and secondary cerebral palsy Kernicterus following prolonged jaundice after birth may result in two syndromes One is an asymmetrical mixed type of cerebral palsy involving pyramidal features but in which rigidity is most prominent secondly a hypotonic child in whom spasticity is not evident

After birth causes include infections such as whooping cough before six months of age and encephalitis at any age Injuries may of course result in sufficient cerebral damage to interfere with neuromuscular function. Convulsions and dehydration in early months of life may result in cerebral hemorrhage or thrombosis Schwartz has shown that areas drained by tributaries of the great vein of Galen are affected by injuries to this vein with secondary hemorrhages

Early Diagnosis

The symptoms of neuromuscular disability are often not prominent enough in the early months of life to bring up the question By six months of age however most cases should be diagnosed. Stiffness in an extremity failure to hold the head up or sit or preservation of a characteristic posture may call attention to the fault.

The baby presented to the physician with the question of possible disability may have the signs elicited by careful examination Over reactivity to a sudden stimulus such as clapping the hands may be present A tendency to hyperextend the neck facial grimaces and tongue rolling may be indicative Observation of the child's activity may reveal quick coordinated motions of one extremity and slow incoordinate motions on the contralateral side It is obvious that allowance must be made for the normal variations and tendency to persistence of hyperactive reflexes in some otherwise normal child

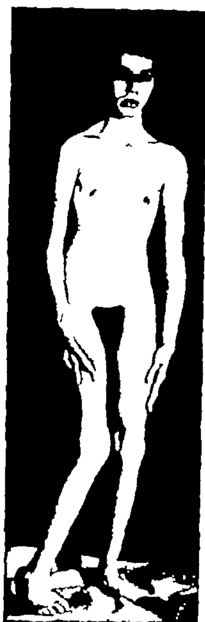


Figure 467 Motor system defect in cerebral palsy increased by the addition of deformity

Most helpful is an inability to shake an extremity into floppy accompaniment to the shake in a short period, particularly when the opposite extremity is readily loosened. The lack of balance exhibited by failure to hold up the head or inability to extend the spine is additional confirmatory evidence in the six month age group. Such evidence alone is not sufficient, but results in guarded observation of the child from then on.

Late Diagnosis

Of the various types of cerebral palsy the one most frequently seen is athetosis. Such a state results in uncontrolled motions and habitual distorted positions.



Figures 468, 469. Inability to extend lower extremities and assume upright posture without support.

The excessive activity and purposeless motions limit the tendency to contracture and any given joint may be passively carried through a full range of motion. In a pure athetotic state the reflexes are not increased and the plantar response is negative. Tension develops in an attempt to control the athetosis but muscle power throughout is good and flail muscles are not seen.

The true spastic individual exhibits increased reflexes, positive plantar responses and stretch reflexes in the muscle. In such individuals a stretch reflex is

elicited by carrying the part suddenly through a range of motion. The opposing muscle contracts at the same point in the arc of motion each time and then with continued steady pressure relaxes to allow continuance through the range of motion. A readily stimulated muscle such as the gastrocnemius opposed by an inactive anterior tibial muscle may result in contractures. The true degree of contracture is elicited by first overcoming the stretch reflex—not by mistaking this response for actual contracture Inactive flail or weak opposing muscles are frequently seen in spastics.

Rigidity on the other hand results in a feeling of resistance throughout the arc of motion rather than at a particular point. Such resistance is also experienced in reversing the motion and extending a previously flexed limb It is a feeling very akin to that experienced in attempting to shift gears in a cold car in mid winter

In the intellectual sphere Brenda concludes that two-thirds of these children have impaired intelligence and only one-third are undamaged. Brenda divides cerebral palsy into the following categories

I Little s spastic rigidity (decerebrate rigidity)
II Pyramidal type (monoplegias hemiplegias, diplegias, paraplegias)
III The mixed extra pyramidal pyramidal type (paraplegias with athetosis)
IV The ataxic-atonic cerebellar type.

Little s Disease

Little originally described this condition in 1853 He noted the predominance of prematurity among these cases exhibiting ' tetanic stiffness" Although Little covered the entire field of cerebral palsy his syndrome is perhaps best limited to conditions involving the extremities and trunk with a decerebrate type of rigid ity This rigidity is the same in all planes of moton. The neck and back are also stiff Such children have a short life and a progressive course. Brenda noted cystic degeneration and edema of the central white matter with the cortex relatively unaffected, but cortical lesions, subdural and subarachnoid hemorrhages were also found.

The Pyramidal Type

Hemiplegia in this type resembles the adult form of cerebral loss of motor function There may be odd variations in addition to limb distribution of in volvement. Thus the rotary type muscles may be involved while the flexor-ex tensor group is relatively unaffected This group comprises the true spastics with typical elbow flexion, pronation flexed and ulnar deviated wrist, adducted thumb and flexed fingers in the upper extremity In the lower extremities the adducted leg with equinus of the forefoot and dorsiflexed first toe are typical Where one limb only is involved or the condition is hemiplegic, the mental defect is usu ally slight. This is usually a group which benefits from training and various orthopedic measures including reconstructive surgery

Here also are the bilateral hemiplegics also known as diplegics in which the legs tend to be more involved than the arms. This category of cerebral palsy is not progressive and tends to remain stationary



Figure 470 Foot deformity in patient with hemiplegia

The brain lesions have been found consisting of cortical degeneration. The cortex is the site of an extensive gliosis with the pia thickened and attached to it. The distribution of the lesion varies from part of one lobe to an entire hemisphere. Possibly venous or sinus thrombosis may be the cause

The Mixed Extrapyramidal Pyramidal Type

This type is slow to become evident and only the failure to achieve a milestone in motor development may first call attention to the difficulty Convulsions may be evident in infancy Spasticity of the lower extremities and uncoordinated flailing flexion-extension motions of the upper extremities may call attention to the athetosis which is present. The head may be noted to be small in the older child.

Venous damage, particularly the vulnerable great vein of Galen with secondary damage through its tributaries apparently plays a large part in this type of cerebral palsy Areas of cortical atrophy and sclerosis beneath enlarged and sometimes thrombotic subarachnoid veins are found The perivascular region of veins running through the white matter is demyelinated and necrotic Subependymal cysts form which lead to degeneration of surrounding tissue

The Ataxic Atonic Cerebellar Type

Here the pathology appears limited to the cerebellum and brain stem with the cerebrum spared and intelligence relatively unaffected. Hypotonic musculature is noted with very late walking as a predominant accompaniment. For a few walking is never achieved. All motions exhibit an unsteadiness and ataxia which is characteristic Aphonia is a prominent feature, but the child often appears to understand well.



Figure 471 Inability to extend wrist in hemiplegia.

Treatment

The cerebral palsy patient presents a multi faceted problem in treatment. Treatment includes understanding the parents the general condition of the patient speech and mental defects convulsions and the motor disability While each of these features demands special understanding and training, the parents seek stability in some one individual in the team usually the one who controls the major phase of the treatment. The fact that many skills are needed in the care of cerebral palsy should not result in the team making a mass approach

From the orthopedic point of view the overcoming of the motor defect is of particular importance

The approach is to the next logical motor accomplishment. The child in order to walk must first learn to sit. The features of the patient that prevent him from achieving his next immediate goal must be overcome be they deformity, contracture ataxia incoordination flail or weak musculature or inability to control motions.

Exercises

Exercises may be divided into two general types The first is passive stretching as a means of combatting contracture, and secondly active repetition of motor acts In using active repetitive motion the position of the limb, the confusion reflex and rhythmic accompaniment may all be used to achieve the desired result. The conversion of exercise into a game-like activity by whatever device increases the ease with which it is done and the duration of doing it.

Braces

Braces which passively hold the patient encased are of little value. The patient cannot exercise within its confines and fails to achieve greater coordination. The bracing problem is quite different from that in poliomyelitis Wherever possible the brace should aid in accomplishing a motion, not in preventing it. In general the most distal major joint is that under poorest control and most in need of help Particularly helpful at the ankle joint is a spring dorsiflexion brace. This is true when the common problem of an overactive gastrocnemius and an anterior tibial inactive in the presence of gastrocnemius tension exists Once an aid to dorsiflexion is incorporated in the gait the patient can be taught to flex the knee and exaggerate the steppage element in the gait thereby improving its character The anterior tibial no longer overstretched re-enters the gait pattern, and each step the patient takes becomes a well controlled one This repeated over the years results finally in elimination of the need for the active bracing.

In the presence of contracting tendencies as in adducted hips flexed thighs, equinus feet, palmar flexed hands and adducted thumbs night casts or splints may be indicated. In sleep, however the patient is usually in a relaxed position

Contracture

The tendency to contracture seen most often in the true spastic or pyramidal group varies from individual to individual It usually requires a constantly tonic muscle excited by minimal stimulus and a flail antagonist or one handicapped by poor mechanical advantage.

Persistent Equinus

The measures to combat this position include passive stretching exercises bringing the foot into dorsiflexion and inversion with the knee extended Wedging casts followed by dynamic splinting usually results in recovery of anterior tibial function and maintenance of the foot in good position Heel cord lengthening has not proven as satisfactory to us as conservative measures. Neurectomy to weaken the gastrocnemius falls in the same category Although in some cases heel cord lengthening may be necessary, it should be carefully evaluated as the gastrocnemius is greatly depended on by the child for stability at both foot and knee. Strayer's method of gastrocnemius recession at junction of tendon and muscle has proven of more value.

Flexed Knees

In this area surgery is most beneficial The attempt to cure the erect posture fault of standing with flexed knees by progressive resistance exercises is in the



Figure 472 Spring brace for dorsiflexion in cerebral palsy a high shoe is necessary in younger patients.

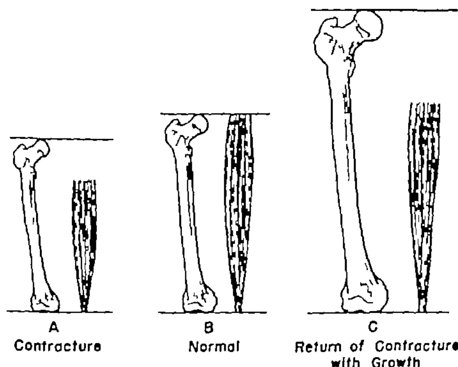


Figure 473 (A) Relative shortness of soft tissues in relation to bone in a contracture (B) Correction to normal situation (C) Return of relative shortening of soft tissues with growth.

majority of instances doomed to failure The quadriceps function is inhibited by the mechanical fault of an overstretched patellar tendon so that the force of the contraction is not transmitted to the tibia throughout the range of motion. Passively the full range of knee extension should be possible and the hamstrings must be weakened to the point of being one grade below the quadriceps. Good quadriceps strength is necessary as well as previous correction of deformity at the hip and knee. A strong gastrocnemius is also necessary These factors were originally brought out by Chandler and re-emphasized by Roberts and Adams. Following this, patellar advancement can be carried out bringing the proximal riding patella several centimeters distal so that it lies in normal position The patella tendon insertion with an accompanying block of bone is moved distal on the anterior tibial surface to accomplish this

Eggers provides for better transmission of the quadriceps force to the tibia by division of the patellar retinacula allowing the patella to ascend and take up the patellar tendon slack from above rather than below

Adduction at the Hip

Weakness of the abductors is a prime factor here and every effort should be made to increase their power rather than weakening the adductors The patient uses them for stability and excessive weakening by lengthening or by obturator neurectomy may result in inability to walk.

Flexion at the Hip

Here again the crux of the problem is to strengthen the glutei Measures which attack the problem from the other side by weakening the flexors must be done with caution. However fasciotomies of the Ober type are occasionally necessary Some patients appear improved by release of flexors such as the rectus femorus as advocated by Duncan.

Pronation of the Feet

Bracing if used, can have an inner T-strap added to aid in holding the foot in inversion. The gradual stretching out of the tight heel cord aids in relieving this constant position. Longitudinal arch pads and inner heel wedges are usually necessary in addition. In this type of foot the peroneal musculature is sometimes found markedly overactive with anterior and posterior tibial musculature not contracting.

A triple arthrodesis is a very satisfactory means of increasing foot strength, stability and function. Such an operation can correct an equinus foot and relieve severe pronation and thereby improve function.

The Limb Alignment

It is hard to get maximum function in the presence of deformity Orthopedic principles of limb alignment hold in cerebral palsy as in other conditions. Excessive valgus at the knee, external torsion of the tibia hyperextension of the knee and foot deformities may all result in limb malalignment which should be corrected.

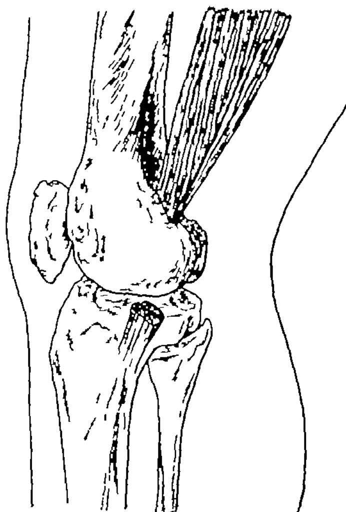


Figure 474 Transplantation of hamstring insertion from tibia to femoral condyle

Upper Extremity

In the upper extremity the most distal joints prove to be those most difficult to control for the patient. Aid to wrist fingers and thumb of a permanent nature enables the patient to then better control the elbow and shoulder

Thumb adduction interfering with the patient's use of the palm may be treated by severance of the nerve to the thumb adductor Fusion of the wrist deviating it dorsalward and ulnarward also gets the thumb out of the palm if the fusion is indicated for other reasons as well.

The palmar flexion and pronation of the hand presents a troublesome problem in many patients. Green has suggested a transfer of the flexor carpi ulnaris tendon from its insertion to the extensor carpi radialis longus tendon. This removes a deforming force and converts the muscle to a supinator as well as a dorsiflexor Such a transplant works well providing there is no loss of kinesthetic sense in the hand.

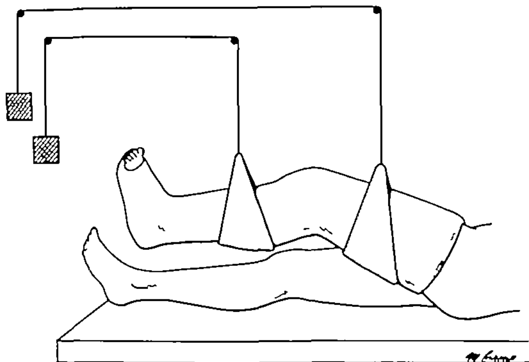


Figure 475 Elevation of the post-operative limb in plaster aids in controlling post operative edema.

Figure 476 Transplantation of the flexor carpi ulnaris (A) to the long radial extensor (B) detail of insertion of tendon into tendon (C)

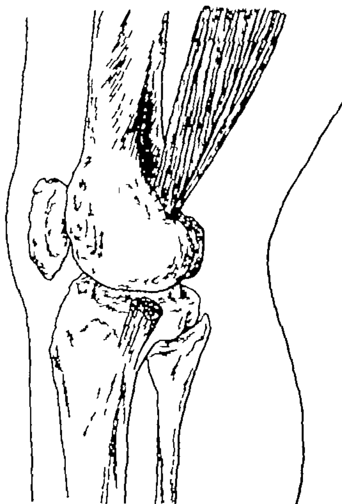


Figure 474 Transplantation of hamstring insertion from tibia to femoral condyle

Upper Extremity

In the upper extremity the most distal joints prove to be those most difficult to control for the patient. Aid to wrist, fingers and thumb of a permanent nature enables the patient to then better control the elbow and shoulder

Thumb adduction interfering with the patient s use of the palm may be treated by severance of the nerve to the thumb adductor Fusion of the wrist deviating it dorsalward and ulnarward also gets the thumb out of the palm if the fusion is indicated for other reasons as well.

The palmar flexion and pronation of the hand presents a troublesome problem in many patients. Green has suggested a transfer of the flexor carpi ulnaris tendon from its insertion to the extensor carpi radialis longus tendon. This removes a deforming force and converts the muscle to a supinator as well as a dorsiflexor Such a transplant works well providing there is no loss of kinesthetic sense in the hand

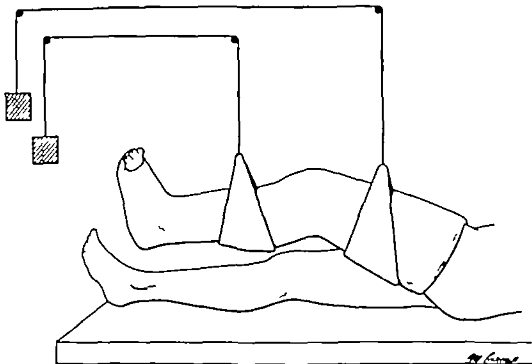


Figure 475 Elevation of the post-operative limb in plaster aids in controlling post operative edema

Figure 476 Transplantation of the flexor carpi ulnaris (A) to the long radial extensor (B) detail of insertion of tendon into tendon (C)

Drugs

Phelps reporting on the use of prostigmine noted that it facilitated correction of contractures particularly in patients with rigidity, apparently lessened the stretch reflex in true spastics and was very helpful in clearing speech difficulties in spasticity or rigidity of the tongue. No effect was noted on pure athetosis. Curare and synthetic derivatives have been used in many centers over the years with disappointing results.

Anti-convulsants drugs are not the province of this text.

References

BRENDA, C E. Developmental Disorders of Mentation and Cerebral Palsies. Grune and Stratton, New York 1952

CLIFFORD, S. H The effects of asphyxia on the newborn infant. J Pediat., 18: 567 1941

EGGERS, G W N Surgical division of the patella retinacula to improve extension of the knee joint in cerebral spastic paralysis. J Bone & Joint Surg., 32A: 80 1950

GREEN W T Tendon transplantation of the flexor carpi ulnaris for pronation-flexion deformity of the wrist. Surg Gynec. & Obst., 75: 337 1942.

GREEN W T AND McDERMOTT L. T Operative treatment of cerebral palsy of spastic type. J A. M A 118: 434 1942

LITTLE, W J Disorders of the Human Frame London, 1853.

McCARROLL, H R. AND SCHWARTZMAN J K. Spastic paralysis and allied disorders. J Bone & Joint Surg 25: 745 1943

PHELPS, W M Description and differentiation of types of cerebral palsy New Child., 8: 1 1949

PHELPS, W M Recent significant trends in the care of cerebral palsy South. M. J 39: 132 1946.

ROBERTS W M AND ADAMS J P The patellar advancement operations in cerebral palsy J Bone & Joint Surg., 35A: 958, 1953.

SMITH C A. Physiological mechanisms of fetal anoxia at birth and cyanosis in the new born. Am J Dis. Child., 79: 1 1950

POLIOMYELITIS

While the diagnosis of poliomyelitis frequently falls on the shoulders of the medical and pediatric practitioner the treatment of its affects is the responsibility of orthopedic surgery A thorough knowledge of the disease the requisites for maximum recovery the prevention of deformity and finally the methods of reconstruction give the orthopedic surgeon tools of great use in understanding and treating other conditions in childhood

The disease may produce an asymmetrical flaccid irregular paralysis without sensory involvement. Such an effect is duplicated by very few conditions.

Past History

Ghormley states that the first monograph on poliomyelitis was published in 1840 by Jacob Heine of Connstatt. Heine put the age group at six to thirty six months and noted that the patients were enjoying unusually good health when stricken Pain and paralysis of both lower extremities was forecast as the result.

Caverly may have been the first to describe the non paralytic form of polio-

yelitis when he reported an epidemic in Vermont in 1894 There were one hundred and ten patients noted to have had paralysis in some form, and at least fifty of these had fully recovered

The first great epidemic appeared in 1905 in Norway and Sweden In this epidemic at eighteen months following onset, fifty six per cent of the five hundred and fifty patients originally reported paralyzed still had some residual paralysis and forty four per cent had recovered completely

In a monograph on poliomyelitis in 1917, Rohrah and Mayer harked back to Badham in 1835 as initiating the use of heat for comfort They mention wrapping the extremities in flannel and supplying heat in the form of hot water bottles

Peabody Draper and Dochez in 1913 divided the types of the disease into the following classifications (1) abortive cases, (2) cerebral group (spastic paralysis evident) (3) bulbo-spinal group (flaccid paralysis) They remarked that preceding the paralysis there is an interval marked by prodromal symptoms and recommended isolation and quarantine at this stage.

Ghormley in his article on the history of the treatment of poliomyelitis brings out their recommendations for treatment. The use of heat, passive motion and rhythmic motion was recommended along with active muscle training to prevent contractures and speed recovery Placing the children in a warm bath was helpful in aiding them to exercise their limbs

Draper emphasized that the resistance to anterior flexion of the spine was more than the stiff neck or Kernig s sign of meningeal irritation and was quite typical for poliomyelitis

Vulpian in the same general era remarked the muscle contracture and deformity appearing following the initial stage of the disease In total paralysis he noted the relaxation of the joint capsule and the tendency for the limb to adapt itself to postural tendencies controlled by gravity The shortening of active muscle groups with development of contracture was responsible for deformity in partial paralysis. The involved muscles undergo marked atrophy, decrease in size and the atrophied fibers become yellowish white due to fatty degeneration Vulpian recommended wet or dry hot packs (1912)

Lovett was a keen student of poliomyelitis and laid the basic groundwork for much of the understanding of the disease as it is today (1917) To Lovett, poliomyelitis was a generalized disease acting on viscera as well as the central nervous system. Spinal cord pathology could produce a fatality via the nerve cells controlling respiration or allowing complete recovery via spontaneous repair He described the symptoms as follows

> The symptoms are in general those of an acute infection In many instances, however gastro-intestinal symptoms predominate while in others, those referable to the respiratory tract are the most marked. Stiffness of the neck to flexion of the head sweating, marked nervous irritability and general hyperesthesia are present in many instances before the onset of paralysis, but they are not all constant

Lovett felt that muscle training forms the basis of the modern treatment of poliomyelitis. In theory it consists of an attempt to make the patient send a voluntary impulse to contract a muscle and of aiding the muscle in contraction

by passive movement by placing the limb in such a position that gravity aids in performing the movement and by the assistance of the hand.

There is one very important point in this connection that is often lost sight of In an affected limb some muscles are as a rule paralyzed, some weakened, and some comparatively normal The muscles that we wish to exercise are the muscles that are paralyzed or weakened. If the exercises are not carefully controlled and located the patient is sure to use strong muscles instead of the weak ones and to develop these and thus to make the muscular imbalance still worse. Loosely given exercises by untrained persons and encouragement to the child to kick around and do anything he can are likely to do harm rather than good.

He emphasized that splints to prevent deformity are an aid to treatment.

Lovett gave us a system of muscle grading which is in common use in following the patient's progress today

The Diagnosis of Poliomyelitis

Despite everyone s familiarity with the cardinal signs of poliomyelitis it continues to be a much misdiagnosed disease. The summer and fall months find virtually every child with a limp stiff neck or pain in the extremities suspected of the disease The problem is often to separate out the cases of multiple neuritis (Guillain Barre), meningitis cord tumor disease of the hip, knee and spine from the cases definitely Heine-Medin s disease or poliomyelitis.

Onset

The onset is like that of many another acute illness. Fever malaise symptoms referable to the gastro-intestinal tract *i.e* vomiting headache, and signs of a mild coryza may all be present. The early period of mild illness may be followed by a week in which the patient feels essentially well This results in a double humped febrile curve since the fever returns followed rapidly by signs of menin geal irritation. A more continuous onset is quite common. The fever does not subside and gradually the symptoms and signs widen to include the central nervous system. It may be that the initial symptoms were overlooked because of their mildness.

In the second phase of the onset muscle aches are common in the extremities. Stiffness of the back and neck are noted, and headaches become a frequent complaint. Irritability common to central nervous system involvement becomes prominent.

Paralysis arises usually in the second to fifth day of the febrile period. It may be maximal in two to four days after recognition. The subsidence of the fever has been felt by many observers to signify the end of development of paralysis. Sensitivity may cause muscles to belie this statement. Marked persistence of the fever may occur in these cases which eventually lead to fatal termination.

As Caverly noted no paralysis at all may develop Sensitivity precedes paralysis. In infants the response may be one of flaccidity such that muscle spasm and sensitivity are not appreciated by the examiner Fever is usually around 101 rising occasionally to 103 but is rarely over this An exceptionally high fever is associated ordinarily with the encephalitic type.

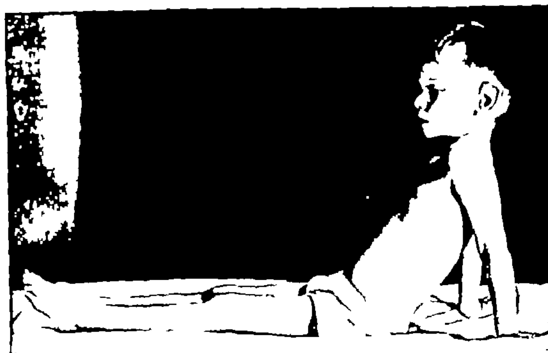


Figure 5. The tripod sign in acute poliomyelitis. Spasm of the hamstrings and back muscles limit the patient's ability to sit up

Clinical Picture

The patient is a very irritable child who resents attempts to handle or move the extremities Unless encephalitic the patient is alert and clear Spasm of the back neck and hamstrings is usual Forward flexion of the neck is painful (Brudzinski) There is a positive Kernig test. Lying on the side the patient does not allow forward flexion of the spine When the patient sits he presents the so-called tripod sign of poliomyelitis With legs extended on the table, attempted sitting is limited and the patient supports himself in partial sitting posture with his arms Straight leg raising is limited usually in a quite symmetrical fashion and not necessarily exhibiting more limitation in the leg which will later show most involvement

Muscles may be sensitive to pressure Muscle sensitivity should be distinguished from nerve sensitivity by palpating the anatomic distribution of the large nerve trunks Spasm of the trunk muscles may be acute anteriorly as well as posteriorly and result in confusion with intra abdominal conditions. Even in the so-called non paralytic type of poliomyelitis some mild weakness may be picked up on careful examination

The Differential Diagnosis

As brought out by Britt Christie and Batson the problem in diagnosis in a medical community alert to the disease is not that poliomyelitis is overlooked but other entities labeled poliomyelitis must be separated out. Other entities to be considered in the non paralytic group are meningitis and encephalitis brain

tumor, gastroenteritis, bacillary dysentery, meningococcemia tick typhus, fecal impaction and intussusception

In those presenting paralysis or pseudo-paralysis one finds the Guillain-Barre syndrome, brain and cord tumors osteomyelitis, pyelonephritis, scurvy, and fractures

The Stages of Poliomyelitis

The stages have been divided into

(1) Acute Stage
(2) The Convalescent Stage
 (a) With Sensitivity Present
 (b) Without Sensitivity
(3) The Chronic Stage

The acute stage has already been described

The convalescent stage starts with the subsidence of fever and continues for a year and a half following onset. During this period it has been found that the disease has a marked tendency for spontaneous improvement in so far as paralytic involvement is concerned. With the appearance of paralysis and sensitivity there is a marked tendency to deformity The paralyzed muscles when opposed by muscles in spasm, become overstretched. Spastic paralysis has been seen very *rarely in encephalitic types of involvement. Flaccid paralysis is the general rule.*

The bulbar and spinal respiratory type need special emphasis since such cases may go on to a fatality Their prompt and early recognition is essential to their treatment. Facial nerves may be involved in bulbar poliomyelitis.

Examination in the stage of sensitivity must be gentle Much can be learned merely from observing the patient's characteristic position in bed Tendencies such as external rotation of the lower extremity drop foot and adduction of the upper extremity are readily appreciated. Assessment of muscle weakness at this initial stage may be general rather than specific but should be sufficient to point out measures necessary to block the development of deformity

Muscle spasm in poliomyelitis is largely that due to an attempt on the part of the patient to protect sensitive muscles Spasm of this type may tend to indicate more paralysis than actually exists since the opposing muscles may be reflexly inhibited. The duration of the paralysis is variable and depends on the degree of involvement. Mild involvement may result in transient weakness only The degree of initial paralysis is of some help in prognosticating the eventual recovery The subsidence of sensitivity greatly aids the patient's progress The recovery curve is greatest in the early phases of the disease and decreases its rate of progress as the end stages of the convalescent period are reached. The patient at the end of this period passes to the chronic or reconstructive stage of poliomyelitis

Pathology

Areas other than the central nervous system are involved. Lymphoid tissue generally is hyperplastic. A myocarditis has been noted at autopsy and occurs in approximately twenty five per cent of cases having a fatal termination. The

nerve cells, themselves, are the seat of primary injury, however The virus is located within the neurons themselves and results in degeneration with secondary tract and nerve degeneration There is an inflammatory and hemorrhagic reaction the subsidence of which in the presence of undamaged ganglion cells may account for the 'recovery tendency' of the disease

The ganglion cells undergo coagulative necrosis, chromatolysis, and neuronophagia There is no evidence that damaged ganglion cells recover The motor cells of the cord are apparently more susceptible than cells of the cerebral cortex. The effects of the disease are not limited to the cord, however, but have been noted in medulla, pons, midbrain, cortex and cerebellum as well. A neuroglial proliferation tends to produce a scar in three to four weeks

There is a tendency for typical distribution of the lesions in the anterior horn, i.e predominantly dorsomedial This accounts for the clinical picture to follow certain distributions of paralysis The anterior horn involvement is most severe, and lateral columns and posterior horns are affected.

In the late stages of the disease the peripheral nerves undergo degeneration of their axis cylinders The muscles undergo the changes associated with denervation atrophy There is a loss of striations reduction in sarcoplasm and alteration in electrical response This is followed by atrophy, fibrosis and fat replacement Bone atrophy is a late development with disuse of the limb

The development of cold legs with tendency to appear mildly cyanotic may be due to parasympathetic palsy from bulbar cord damage with sympathetic system spared through the lateral columns of lumbar one and two Anterior horn cell damage affects the parasympathetic system more than the sympathetic resulting in failure of bladder and rectum to contract and thereby produces the occasional clinical problems in these areas seen in poliomyelitis

Treatment

In the acute phase the patient is on bed rest with general measures including positioning to guard against deformity This includes the use of a bed board so that excessive lumbar lordosis is readily seen. The bed board should not be one piece, however since treatment of the disease includes the periodic positioning of the patient in gentle spine flexion when mobilization is begun. A knee roll to allow slight knee flexion is a matter of great comfort to the patient The foot board is separated from the end of the mattress with the heels allowed to drop into the interspace. A small neck roll may be of comfort to the patient. Hot packs are used to diminish sensitivity and to aid the performance of stretching type exercises by being used just prior to the stretching. If they produce discomfort, they have little value. Hot packs in general are wrapped around an extremity and laid on the trunk. Continuous use of packs can be debilitating to the patient.

There are many special therapeutic problems at this stage which cannot be covered here They include measures to relieve difficulty in swallowing respiration, and micturition The use of the respirator rocking bed and tracheotomy have special and definite indications

While the patient is febrile it is important that disturbances and handling



Figure 478 Bracing is a means of expanding the child's horizon—not limiting it

are minimized and that supportive measures be gentle and adequate Positioning in bed to avoid deformity will result in a very sensitive patient if it over stretches irritable muscles

Once afebrile, an estimation of the paralytic involvement is obtained. This may be inaccurate due to muscle spasm and sensitivity and indicate more severe involvement than actually exists Deforming tendencies are noted and the patient positioned against them periodically during the day The bed may be used to aid in developing spine flexion and hamstring stretching No positioning should be considered static but changes in position should be frequently performed

Splints are often necessary In younger children they are used to substitute for the foot board since difficulty is often found in keeping the youngster against it. The splint holds the foot in neutral so far as dorsiflexion and inversion eversion are concerned An out-rigger attached to the splint to prevent chronic hyper extension of the knee by holding it in slight flexion is occasionally used. Splints should not be misused by being left on the patient for excessively long intervals

The patients should have all joints carried through a range of motion within the limits of pain, regularly during the day Stretching is begun early and carried to but not beyond the point producing pain. Limbs are held at bony prominences not in areas of large muscle masses in tender patients The period of convalescence is greatly shortened by early stretching and mobilization The spine should be stretched in rotation as well as in flexion.

The drop foot deformity in poliomyelitis is well known. When the patient is prone, the feet should be allowed to drop into the interval between the mattress and the foot board. Less well known but quite common is the development of a flexion abduction deformity at the hip This deformity gets its start with spasm of the ilio-psoas, sartorius and tensor fascia lata muscles which tilt the pelvis forward producing excessive lumbar lordosis. When the spasm is unilateral, a curve in the spine is produced. Full spine roentgenograms in recumbency are often helpful in picking up early tendencies to spinal deformities.

By the end of the first month the patient ordinarily has full straight leg raising and spine flexion and an accurate estimation of the paralysis has been obtained. The following muscle grading system is usually used

MUSCLE RATING	DEFINITION
0	No palpable contraction
Trace	Palpable contraction but no movement of the part
Poor	Moves part through its range but not against gravity
Fair	Moves part through its range against gravity but not resistance
Good	Moves part through its range against gravity and against against resistance
Normal	Normal

In general a muscle rated zero cannot be expected to progress beyond a poor rating. A muscle needs to be rated as fair in the early stage of the disease in order to develop to good or normal in most instances. Not the extent but the degree of involvement is of some help in prognosis. Certain muscles have a

marked tendency to involvement together i.e., biceps femoris and gastrocnemius, and the clinical problem can often be predicted when the involvement of one is known

The basic exercise rule is to exercise the muscle in its rated grade. Exercising the muscle below what it can do will not produce improvement except in endurance In order to procure increase in power the muscle must perform maximum work at each contraction Increase in power is more helpful to the poliomyelitis patient at this stage of his disease than increases in endurance. Repetitive exercises to increase endurance may be added at a late date. When the limb can be moved against gravity progressive resistance exercises are added

In general the patient is kept in bed *when he has something to gain from so* doing. Residual sensitivity or tightness should be overcome before the patient is allowed up Asymmetrical tightness in the trunk should not be allowed to persist Weakness of the trunk often calls for continued bed rest. The development of an extremely important muscle such as the gastrocnemius gluteus medius or gluteus maximus may rate further care before ambulation.

With the exception of non paralytic cases, crutches are almost a routine when starting ambulation Even in the abortive cases the child may have difficulty in raising the body weight with the gastrocnemius and crutches and heel lifts are a help in procuring return of full function of the gastrocnemius within two weeks of being allowed up

Bracing is a means of preventing deformity and allowing greater activity than would otherwise be possible Many of these children can walk without braces but with severe deforming tendencies in their gait A brace aids in the prevention of overstretching of a muscle. Overstretching may result in regression rather than in an increase in the power of a muscle. The gastrocnemius is such an important muscle in a child s gait that it must be protected at all costs. The young child without the gastrocnemius will walk with hyperextended knees and frequently needs a long leg brace The older child can be taught to keep the knee flexed if sufficient muscle power is present.

The weak gastrocnemius must be prevented from allowing a calcaneus foot to develop The particular type of brace used varies from child to child depending on the individual muscle picture

Paralytic shoulders (weak deltoid) drop feet(weak anterior tibial) knee weakness(quadriceps or hamstrings) are among other areas that must be protected by apparatus. Trunk weakness must be braced depending on the degree of involvement. Abdominals are particularly prone to being overstretched on resuming the upright posture.

The pool is of particular use in exercising patients with trunk and hip weakness in handling large patients with extensive involvement, in stretching out resistant tightness and in starting ambulation in patients with minimal musculature.

So far as electrical stimulation is concerned the present evidence indicates that in the initial phase of involvement it temporarily slows the progress of atrophy However atrophy will progress none the less and eventually reach the same point that an unstimulated muscle similarly involved would reach. Cessa

tion of the stimulation results in rapid progress to reach the level of atophy expected There appears to be no justification for its routine use It may be of some help in enabling the patient to more readily find a muscle as in facial paralysis or in virtually flail extremities

CHRONIC OR RESIDUAL POLIOMYELITIS

After the convalescent period of poliomyelitis is over, that is, eighteen months after onset the patient enters a period dominated by several important principles The first of these is prevention of deformity, the second is increasing muscle power, and the third is reconstruction and substitution for paralysis

Although defined as a period in which no further recovery can be expected this does not rule out the ability of the patient to gain in the performance of practical functional tasks

Clinical Description

These patients have usually recovered from the initial mental depression in poliomyelitis They enter the residual stage attempting to gain as much increased functional ability as possible Except in the case of a completely flail extremity they are left with muscle imbalance This alone and unprotected is sufficient to cause deformity To this imbalance may be added the additional deforming tendencies due to use and due to gravity Atrophy of the extremity noted within six weeks of onset is present The circulation may be markedly diminished and in childhood will have an effect on growth The patient up to this point has not been allowed to substitute muscles to gain function and has become well schooled in exercise routines The economic factor an important consideration in adults, is not active with children.

Management

This stage may be attended with loss of function due to progressive deformity Flaccid paralysis is sufficient disability but this may be increased by contracture Gravity and function may stretch ligaments and joint capsules to the point where they no longer act as adequately functioning units

Bracing

The brace is a means of allowing increased activity without increasing deformity It is not a means of limiting activity Bracing should be as dynamic as possible The following outline is greatly simplified

Lower Extremities

The foot, if completely flail must be supported on both sides and anteriorly and posteriorly by a double upright brace with stops to limit both dorsal and plantar flexion. Such a brace ordinarily allows ten degrees of motion at the ankle as an aid in walking.

When the peroneal musculature alone is weak a single upright brace may be sufficient placed on the medial side. An anterior T-strap is ordinarily added This strap runs from the shoe to the lower tibia and upright. Lateral wedges



Figure 478 Calcaneous foot produced by attempting take off in the gait without gastrocnemius power

may be used on the shoe. Stops are added when there is weakness antero-posteriorly An anterior stop aids in preventing overstretching of the gastrocnemius. A posterior stop aids in preventing drop foot and overstretching of the anterior tibial.

Anterior or posterior tibial weakness is protected by a lateral single upright with an inner T-strap Medial wedges on the shoe and a longitudinal arch pad are often necessary in addition. Anterior or posterior stops or both may be necessary

Every contingency cannot be covered and different combinations of paralysis may demand double upright braces where at first glance a single upright may seem to be all that is necessary

The Knee

A poor or worse rating of the gastrocnemius may result in a tendency to hyperextend the knee despite good knee musculature. In children age six and under a doubt in regard to the necessity for a long leg brace should result in its use. Older children can be taught to protect their knee, by walking in flexion.

Surprisingly both strength of the quadriceps in the presence of weak hamstrings and weakness of the quadriceps may result in hyperextension at the knee A brace to support the knee must of necessity be of double upright construction. The leather knee cap should not be so tight that it forces the knee into hyperextension Knee braces have a number of ingenious locks designed to allow flexion when sitting. The single so-called drop-lock works well in children.

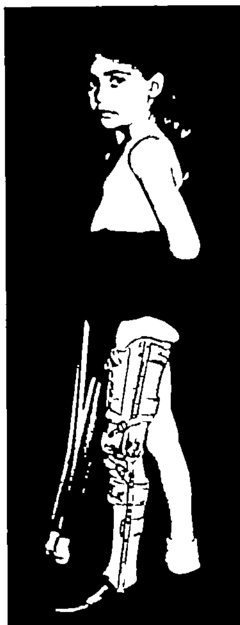


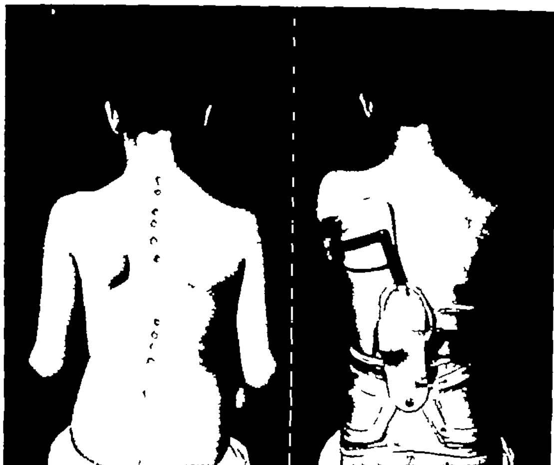
Figure 480 Long leg brace for flail leg.

The Hip

Gluteus medius and maximus limp are largely eliminated by the use of crutches or canes The hip weakness cannot be substituted for by a brace alone. Where the hip is flail it may be necessary to joint a long leg brace to a trunk brace by means of a hinge

The Trunk

Where abdominal weakness alone is present and is relatively symmetrical a corset alone may suffice. The corset should be laced from below upward to properly support the abdominal contents. Lateral deviations of the spine may be well held by a brace of the Barr Buschenfeldt type with the arms of the brace appropriately placed to eliminate the deviation In general the lower arm sup-



Figures 481 482 Barr Buschenfeldt brace used to hold early paralytic scoliosis

ports the slump The upper arm corrects the curve. Check roentgenograms holding the trunk with the hands in the position of the arms of the brace may be taken. An x ray film taken in the brace will reveal the need for further adjustment if necessary

Neck

Asymmetrical weakness of the sternocleidal mastoids may call for holding braces of the Buchminster Brown or collar type in a growing child. Where the weakness is quite general braces built with chin cups and occipital supports deriving their support from a pelvic band in the fashion of Blount s brace may be needed.

Upper Extremity

The shoulder joint is subject to gravity and weakness of the deltoid may result in a tendency for overstretching of the capsular structures. Support for the humerus may come from the trunk or via a sling anchored at the neck. Allowing a subluxation to develop renders more difficult a later reconstruction and eliminates any possibility of improvement.

The flail elbow may be simply kept at a right angle by a sling or by a combination shoulder and elbow brace

Hand bracing is an individual problem in which the position of function is encouraged and has been more fully described elsewhere

Deformity

The adult presents a different picture from a child In children growth in length of bone relative to soft tissue is a factor in producing severe contractures. Children may have to be supported many years in order to get sufficient skeletal maturation of a joint to perform an arthrodesis Children wandering from supervision may in a much shorter interval develop more severe deformity than an adult The problems of rehabilitation both from the point of view of surgery and vocation are quite different.

Progressive Resistance Exercise

Exercises during the chronic stage of poliomyelitis may be aimed at development of hypertrophy Progressive resistance exercises are particularly valuable in this regard as popularized by DeLorme and Watkins. The principle used is that to gain strength the muscle must undergo a contraction which is maximal—that is maximal in relation to the work load performed. Thus if a muscle can lift five pounds against gravity, it should be exercised at this level until it can lift six pounds Repetitive submaximal contractions may increase endurance but not strength.

Since a heavy load increases the sensory response in a muscle, progressive resistance exercises aid also in isolating and strengthening transplanted muscle. DeLorme has advocated preoperative use of this type of exercise in order to isolate function The problem of training the muscle in the new position is then greatly simplified. The stages in educating a transplanted muscle involve first the use of the muscle while suppressing the use of antagonists to it. This involves repeated "setting" of muscle until a powerful sustained contraction is reduced. The proprioceptive response aids in finding the muscle when some form of resistance is used The motion of the part involved aids in dropping out the antagonist muscles Secondly the synergists are dropped out one by one leaving the transplanted muscle which once isolated in this manner can progressively develop greater force with increasing resistance

Stretching Exercises

These exercises have their greatest use in combating contractures which play an active role in the development of deformity Many of these contractures are residual from the onset of the disease—a period when muscle spasm resulted in habitual positioning of joints. Myostatic contractures take place only in innervated muscle and are much more likely to be seen in partially involved than in completely flail extremities.

Fixed equinus of the foot due to anterior tibial weakness in the presence of a strong gastrocnemius is most common Flexion contractures of the hip aided by a strong iliopsoas and a weak gluteus maximus are frequently unrecognized till

well marked This is particularly true of a flexion abduction contracture ' the sartorius and tensor fascia lata enter as additional factors.

Contractures of the trunk limiting lateral bend, forward flexion or rot may be factors in the development of trunk deformity

Stretching type exercises must be in the correct position so that the proximal to that being moved is immobilized The knee is extended whe achilles tendon is stretched for example. Correct use of stretching exe requires a detailed knowledge of anatomy

Stretching exercises accomplish their purpose when done to the maximum day rather than when repeated many times in a single day Muscles such a hamstrings are particularly easy to stretch. Those about the pelvis with b fascia and short muscle bellies particularly are difficult.

Functional Training

The simplest example of functional training is the teaching of a patie negotiate stairs. It is obvious that the strong leg pulls the patient upstairs steadies him as he advances downstairs Such a task has to be confronted by patient to be appreciated, however and the bracing and muscle power th patient has must be converted to use in the tasks of life. The simple act of get out of a chair can be a very complicated thing when one is handicapped. combining of crutches and their use as an aid rather than a hindrance ca taught quickly to the patient. Yet if left to his own devices the patient may it impossible to leave the chair with any facility The necessity for arms, the of the upper extremity to extend the trunk the locking of braces in turn bec aids under guidance.

The practical problems that the patient will have to face must be met solved These include eating the use of the bathroom, stairs entering and lea public transportation etc. One need not have expensive equipment to dupli these tasks. Ingenuity and forethought will bring the patient to the succes *solution of these things if they are met in some form in advance of activit*

Reconstruction of the Paralytic

No operation of this type is justified unless it either (1) enables the pat to use less apparatus (2) prevents progressive deformity or (3) increases patient s ability to perform. It is recognized that there are many alterna procedures from the one more or less standard procedure mentioned.

The Unstable Foot

There are various types of situations which have to be met under this head since all situations cannot be covered exactly In various borderline situati individual judgement must enter in. Bony deformity must be corrected. The of muscle transplant alone will be limited in the presence of deformity

I. *Peroneal Strength anterior and posterior tibial weakness and good gastr nemius—result ordinarily in a valgus foot and drop foot*

Below the age of ten the foot has been stabilized from the skeletal point of v by the subtalar bone block procedure of Grice. A transplant alone in the pr

ence of this deformity cannot pull the medial arch up against the body weight. In this procedure the subtalar joint is exposed The os calcis is inverted beneath the astragalus and a notched bed is made in the inferior surface of the astragalus and the superior surface of the os calcis just distal to the posterior third of this joint

The height of the bed is then measured by compass Two grafts from the proximal tibial cortex are then inserted exactly into this bed to block the ability of the os calcis to evert laterally The grafts are wedge-shaped with their greatest height laterally

A transplant of the peroneus longus is then performed The peroneus longus tendon is divided just proximal to the os peronei The peroneus brevis is detached from the base of the fifth metatarsal and sewn to the stump of the longus A bed is then made for the insertion of the tendon in the base of the second metatarsal (There may be feet where a still more medial insertion is indicated) An insertion that has worked well is the creating of a sufficiently large drill hole in the superior cortex to accept the tendon Two small drill holes are made just distal A third incision is now made on the lateral calf The peroneus longus is visualized identified and then retracted into the proximal wound. A long incision is made into the anterior muscle compartment A tendon passer is introduced into this compartment and run beneath the deep fascia to the bed prepared in the



Figure 483 Peroneal strength and anterior and posterior tibial weakness leads to valgus deformity of the foot as on left.

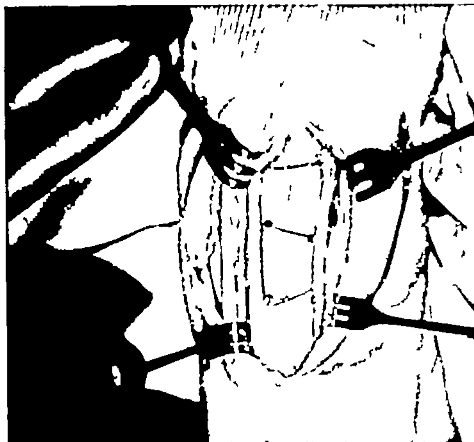


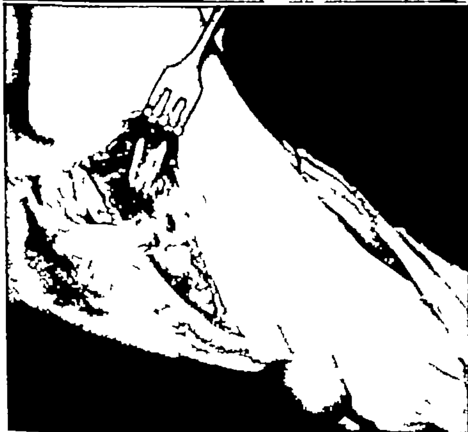
Figure 484 Peroneus longus muscle implanted into base of second metatarsal

base of the second metatarsal. Silk sewn into the peroneus longus is drawn by the tendon passer down this route. If done quickly and smoothly the peroneus longus will lie flat without twisting

Using good tendon suture technique the two ends of the thread (2-0) are passed individually out through the distal drill holes The tendon is drawn down into the bone and the thread tied The tendon should be taut with the foot in maximum dorsiflexion and inversion. The patient is kept in a long leg plaster cast in the corrected position for six weeks and then in a bivalved cast in which the peroneal transplant can be exercised. This situation exists until the trans plant is found and working well When bone union is demonstrated in the grafts, the patient starts weight bearing with crutches. When the use of the transplant in the gait is established the crutches are then discarded (about two months later)

Over the age of ten and in feet with a fixed valgus deformity a triple arthro-





Figures 485, 486 Subastragalar arthrodesis described by Grice (485 above) The tibial cortex is cut on a carefully measured triangle twice the height of block needed with the center cut angled so that grafts can be wedged into subastragular joint (486 below) The tibial cortical grafts in place.

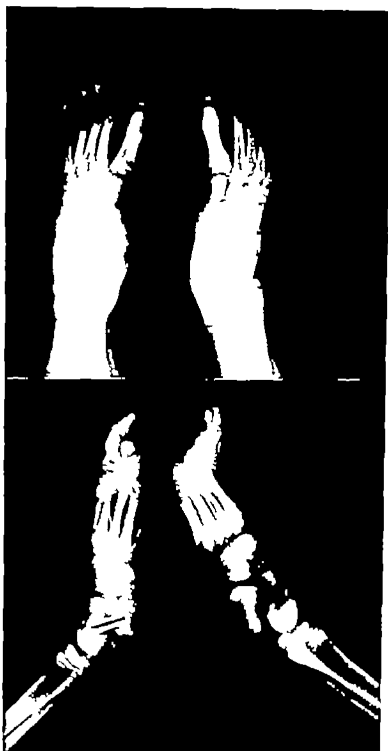


Figure 487 Postoperative roentgenogram of tibial cortical grafts blocking subastragular joint into inversion.

desis is indicated. In addition however, the muscle balance of the foot must be restored by a peroneal transplant or the deformity will tend to recur

A triple arthrodesis corrects bony deformity by the shape of the wedges removed and by fusing the mid tarsal and subtalar joints provides stability for the foot

A lateral incision over the sinus tarsi is made The muscle belly of the short extensor to the toes is reflected from its origin on the os calcis The long extensor tendons are retracted superiorly and the peroneal tendons retracted inferiorly The fat filling the sinus tarsi is removed The head of the astragalus astragalar surface of the navicular, cubo-calcaneal and sub talar joints are exposed. The first osteotome cut is made in the astragalus in line with the coronal plane of the ankle joint and without involving the bone further than is necessary to get in this plane Navicular and cuboid joint surfaces are removed at right angles to the forefoot The sub-talar joint surfaces are removed, astragalar surface parallel with the weight bearing plane of the ankle joint, calcaneal surface with sufficient wedge to correct the hindfoot eversion, but inversion should not be allowed as this will increase with weight bearing

At the conclusion joint surfaces should be flatly opposed The tissues are repaired to eliminate dead space The foot is held by the operator as a long leg plaster case is applied This is kept on for six weeks At this time the cast is removed roentgenograms are taken and a short leg cast applied, if early fusion is apparent The short leg cast is used for two weeks with crutches and a walking heel then applied for two more weeks to stimulate maturation of the fusion The cast is then removed and partial weight bearing with crutches is begun.

When a transplant has been done simultaneously, the cast is bivalved at six weeks and exercises for the transplant are done and continued for a usual four to six week period until maturation of the fusion is revealed by roentgenogram plus action of the transplant. Partial weight bearing with crutches is then begun.

II. *Anterior Tibial absent, strong peroneals, posterior tibial strong strong toe extensors*

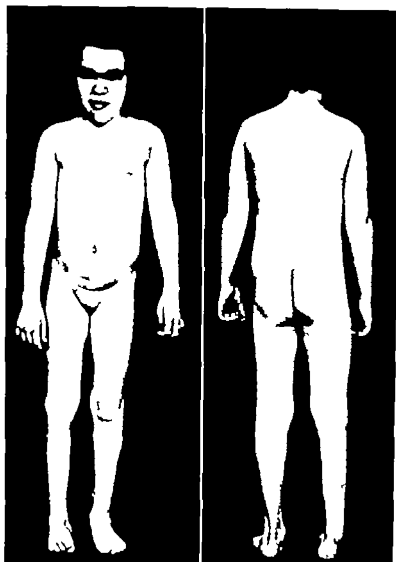
When the posterior tibial balances the everting pull of the peroneals equinus of the forefoot may result in a cavus type deformity Before fixed bony changes occur transplant of the extensor tendons into the metatarsal necks gives dorsi flexion.

III. *Gastrocnemius weakness, strong peroneal and anterior and posterior tibial muscle power*

Below the age of fourteen the peroneal longus posterior tibial and long toe flexor may be moved to insert on the posterior inferior angle of the os calcis. When there is no gastrocnemius power such a transplant, while not giving sufficient power to substitute for a brace, may be sufficient to prevent progression of a calcaneous deformity

When there is some gastrocnemius power, such additions may be sufficient to substitute for a brace and give a normal gait.

Over the age of fourteen an ankle fusion is preferable in the presence of a laterally stable foot.



Figures 488, 489 Inverted foot with poor gastrocnemius, and peroneal power and present anterior and posterior tibial.

An incision is made antero-medially and carried down to the ankle joint between the long extensors of the toes and anterior tibial tendons A Hatt type of procedure is followed. The cartilage surfaces of the ankle joint are denuded and fitted together A graft of tibial cortex is elevated from the antero-medial surface of the distal tibia. The most distal inch of tibial cortex is left intact as a bridge. A Hatt osteotome is inserted into the medullary cavity of the tibia driven across the ankle joint and well into the astragalus. The bone graft is then driven down the track made by the osteotome The foot is very carefully positioned as this is done so that the position determined as optimum pre-operatively can be achieved and held by the graft. Ten degrees of equinus is usually necessary with some variety dependent on sex, type of heel worn, etc

The Knee

Hyperextension Deformity of Knee

The need for correction of this deformity is quite frequent It results from locking the unstable knee as a gait habit by placing it in hyperextension This procedure is not done till growth is completed A Steinman pin or Kirschner wire is driven into the most proximal tibia Force is applied to the pin as the tibia is divided distal to it The proximal fragment is thus held in hyperextension as the distal fragment is lined up with the femur This position is held as a plaster cast is applied

Absent Quadriceps, Strong Semimembranosus Semitendinosus and Biceps Femoris

Not every patient with an absent quadriceps needs muscle power anteriorly at the knee Where there is a fundamental gain possible one or more of the hamstrings may be transplanted forward to the patella

The Hip

The most troublesome situation here is the weak absent gluteals with a strong hip flexor The result is a tendency to hip flexion contracture starting a series of events which may result in a paralytic dislocation of the hip

Barr found anterior fasciotomy at the hip did not prevent a recurrence of deformity with reenforcement posteriorly Such reenforcement is available in the erector spinae transplant of Ober It is insufficient to substitute for a gluteus maximus but can be used as a check to the development of a hip flexion contracture

In this procedure after preliminary fasciotomy anteriorly a second incision is made to detach the fascia lata at its insertion into the fibula It is withdrawn and run from the insertion of the tensor fascia lata into it, through a subperiosteal tunnel below the greater trochanter posteriorly over the pelvis, through the substance of the gluteus maximus to the erector spinae The lower end of the erector spinae is fashioned into a cone in its lower segments and the fascia lata sewn around it with the hip in an extended position.

The Shoulder

Paralysis of the deltoid can be very well substituted for by fusion. There are fundamental conditions to be met, however before fusion can be considered The muscles controlling the scapula must be rated good or better the hand and elbow must be functional. The situation of the other arm must be considered. The patient should be above age twelve The joint is usually denuded of cartilage, surfaces opposed, and the acromion osteotomized and bent down to notch into the humerus

The usual position is such that the hand reaches the mouth by flexing the elbow This usually means about forty five degrees of abduction fifteen degrees of forward flexion and fifteen degrees of internal rotation.

The post-operative shoulder spica is made pre-operatively so that the position

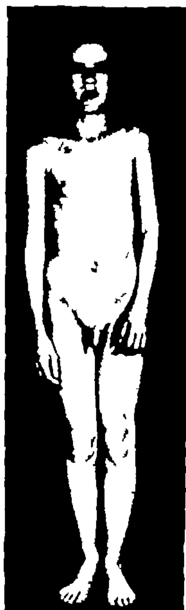


Figure 490 Functional hand and elbow with flail deltoid but good scapular muscles—a candidate for fusion.

will be accurate The spica is removed and replaced after operation. The usual period of necessary immobilization is four months.

The Elbow

Lack of elbow flexion must be overcome to restore useful function to the upper extremity It is fairly common to have a good forearm flexors with the weak or absent biceps. If the hand has function a Steindler transplant may be performed.

Here the common flexor tendon is isolated at its origin from the medial epi condyle and transplanted approximately one and one-half inches proximally up the shaft of the humerus This improves the lever action across the elbow joint so that the hand can be elevated against gravity and with some resistance.

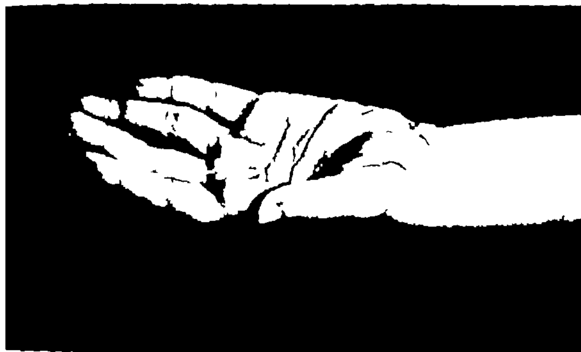


Figure 491 Muscle transplant—in this case 4th sublimus tendon to the metacarpal and first phalanx of the thumb

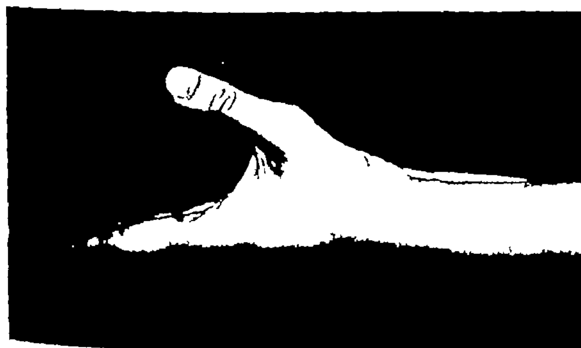


Figure 492 Muscle transplant provides motor power

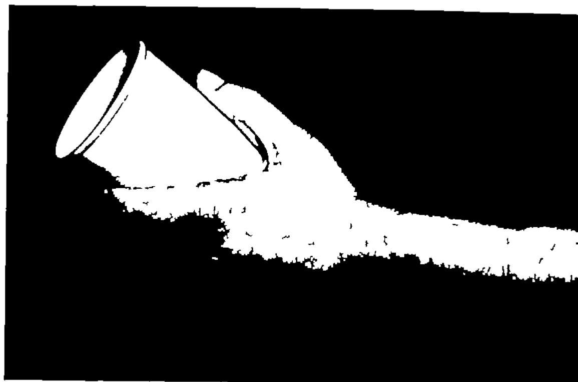


Figure 493 Muscle transplant increases function

References

BOUMAN H D AND SCHWARTZ, R. P The degree the extent and the mechanism of muscle spasm in infantile paralysis. New York State J Med., **44**: 147 1944

COLONNA, P Hamstring transplantation for quadriceps paralysis. J Bone & Joint Surg., **5**: 472, 1923

GRICE, D S Further experience with extra-articular arthrodesis of the subtalar joint. J Bone & Joint Surg., **37A**: 246, 1955

GHORMLEY R. K History of the treatment of poliomyelitis. J Iowa M Soc. **37**: 343, 1947

GREEN W T Diagnostic and therapeutic considerations in anterior poliomyelitis. Rhode Island M J **28**: 89 1945

JOHNSON R W Results of modern methods of treatment of poliomyelitis. J Bone & Joint Surg **27**: 223 1945

JONES R On arthrodesis and tendon transplantation. Brit. M J March 28, 1908.

LEGG A T AND MERRILL, J B Physical therapy in infantile paralysis. W F Prior Inc Hagerstown Md 1932.

LOVETT R W The treatment of infantile paralysis. Blakiston, Philadelphia, 1916

PEABODY C W Tendon transplantation—an end-result study J Bone & Joint Surg., **20**: 193 1938

SABIN A. B Pathology and pathogenesis in human poliomyelitis. J A M A **120**: 506 1942

STEINDLER, A The Rehabilitation of the Paralyzed Patient. Appleton-Century-Crofts, New York 1925

YOUNT C C The role of the tensor fascia femoris in certain deformities of the lower extremities. J Bone & Joint Surg **8**: 171 1926.

PROGRESSIVE MUSCULAR ATROPHY OF THE PERONEAL TYPE

Progressive muscular atrophy of the peroneal type begins in the feet and legs and extends only in later years to the upper extremity. The disease inevitably progresses but patients are found in the sixth and seventh decade of life. Allen in 1938 found an inheritance pattern with three possibilities: dominant, sex linked recessive and simple recessive. He concluded that one could prognosticate from the pattern of inheritance the age of onset and clinical severity.

Rarely the disease can be first noted under the age of one with the vast majority of onsets below the age of twenty. The disease is apparently a primary anterior horn cell atrophy with secondary involvement of the peripheral nerves.

Clinical Picture

The patient often first presents with a foot deformity. The deformity begins with varus of the hindfoot, then the development of equinus, and finally a cavovarus foot with clawing of the toes. There is peroneal muscle weakness followed by involvement of the extensor digitorum longus, anterior tibial and gastrocnemius. The atrophy is symmetrical and peripheral. In the hands the intrinsic musculature, and a symmetrical forearm atrophy appear. It is primarily a motor involvement although occasional areas of hypesthesia and loss of deep sensation are found. With loss of motor power the muscle reflex disappears.

Treatment

Because of its prolonged course such cases are well suited to measures which correct the foot disability. Braces and exercises are an aid but foot stabilization procedures have proven of even greater value. Jacobs and Carr have advocated a foot stabilization preceded by division of the plantar fascia or lengthening of the tendo achilles as indicated. Appropriate wedges of bone are removed as the triple arthrodesis is done correcting the foot deformity. These authors also advocated the transfer of the posterior tibial tendon through the interosseus area to either the cuboid or middle cuneiform as a means of balancing the foot musculature.

References

ALLAN, W. Relation of hereditary pattern to clinical severity as illustrated by peroneal atrophy. Arch. Int. Med., 63: 1123, 1939.

DUNCAN, S. A. Orthopedic treatment of the muscular dystrophies and muscular atrophies. Virginia M. Month. 64: 40, 1937.

HOKE, M. An operation for stabilizing paralytic feet. J. Orthop. Surg. 3: 494, 1921.

JACOBS, J. E. AND CARR, C. R. Progressive muscular atrophy of the peroneal type. J. Bone & Joint Surg. 32A: 27, 1950.

10

Malignant Tumors of Bone

OSTEOGENIC SARCOMA

Osteogenic sarcoma is a malignant metaphyseal tumor primary in bone with a bad prognosis. It occurs in other age groups but the highest incidence is reached between ten and twenty years. It is not the most common of malignant tumors in childhood and fortunately is quite rare The tumor initially metastasizes to the lungs via the blood stream.

Malignant tumors of bone are hard to classify and must be classified on the basis of their entire natural history rather than on the basis of one isolated technique.

Clinical Picture

The outstanding features are first pain and secondly the presence of a mass. The pain is continuous frequently making rest difficult The patient tends to lose weight, and become haggard and drawn The mass may not be immediately evident unless it is in a subcutaneous location. When evident, it is firm and its edges are continuous with the bone structure It is tender and frequently accompanied by dilatation of veins and some increase in local heat in the area. The persistent pain has usually resulted in failure to continue normal use of the limb and considerable muscle atrophy is often present.

Distribution

Osteogenic sarcoma is typically a metaphyseal lesion and very rarely diaphyseal Approximately seventy five per cent are located about the knee in either distal femoral or proximal tibial metaphysis They have also been found in the proximal humerus femur ulnar and fibula as well as distal ulna and fibula. Other locations are exceedingly rare and atypical.

Pathology

There is considerable variation in the gross appearance due to the variation in bone production associated with the tumor An osteogenic sarcoma, however

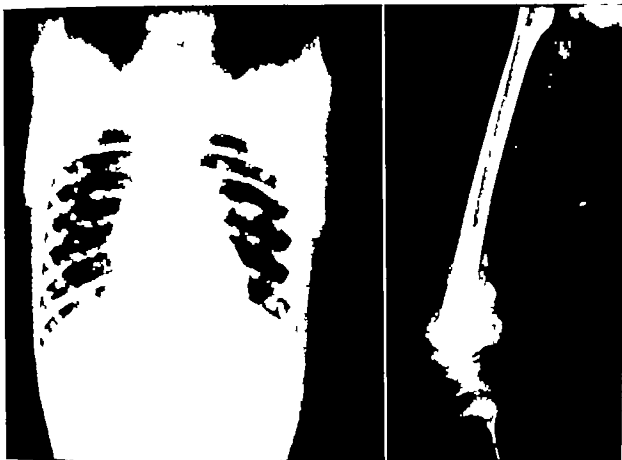


Figure 95 Osteogenic sarcoma of the femoral metaphysis with osteoplastic metastases to the lung

tends to be metaphyseal and to break out of its location in the bone rather than running axially along the medullary canal as does a Ewing's tumor

The area where the cortex is broken through is covered by the tumor mass which is rounded and may be soft or firm There is considerable vascularity

The actual tumor mass may be overlaid by heavy fibrous tissue. Biopsy should include the deep as well as superficial layers and may demand the removal of a block of bone containing not only bone but tumor areas as well

The structure miscroscopically is highly variable with areas of necrosis highly cellular areas and areas of bone formation Although grossly the tumor might be characterized as osteolytic or sclerosing microscopically all types of activity are usually found in various fields The cells have been characterized as osteoblasts and usually show considerable variation in size hyperchromatic nuclei and numerous mitoses Metastases when they occur are to the lung unless in the terminal stages of the disease The lung metastases produce bone or cartilage.

Roentgen Picture

The lesion exhibits active destruction of bone It tends to be rounded rather than linear Areas of gross tumor bone can usually be seen. At the edge of the tumor there are often areas of bone reaction subsiding still further from the

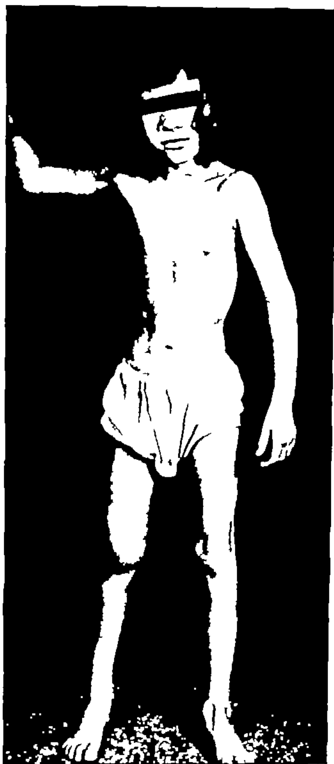


Figure 495 Clinical appearance of patient with osteogenic sarcoma There is swelling of the metaphyseal area of the distal right femur

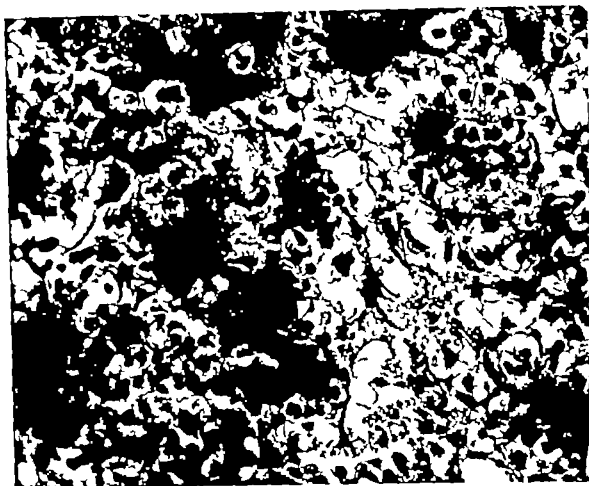


Figure 496 Bone formation in osteogenic sarcoma

tumor into subperiosteal calcification of a stasis type The breaking through of the subperiosteal calcification by the tumor leaves a triangular area which is known as "Codman s triangle" The extension of the tumor tends to produce a sunburst appearance with the areas of ossification radiating from the central originating area.

The involved area is totally destroyed, but not totally radiolucent for it is filled with its own density formed of the tumor tissue itself and tumor ossification.

Treatment

Treatment is preceded by biopsy and examination on permanent nonfrozen sections. The results of treatment are poor but not hopeless Many regard the childhood form of osteogenic sarcoma as the most malignant.

The statistics regarding osteogenic sarcoma reveal that those patients with late amputation (that is amputation after a period of months in which the tumor was known to exist) have a higher survival rate than those with immediate amputation (Ferguson) Whether this reflects only a lower degree of malignancy or

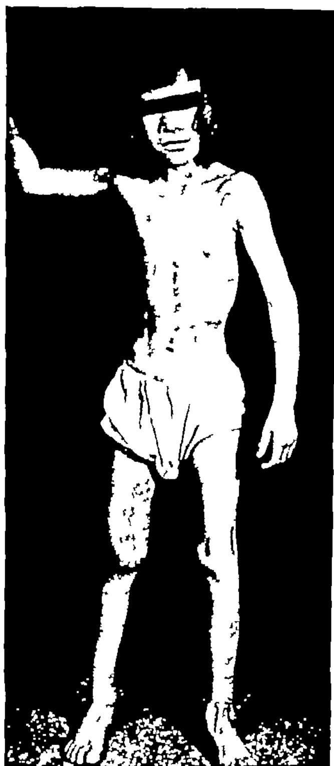


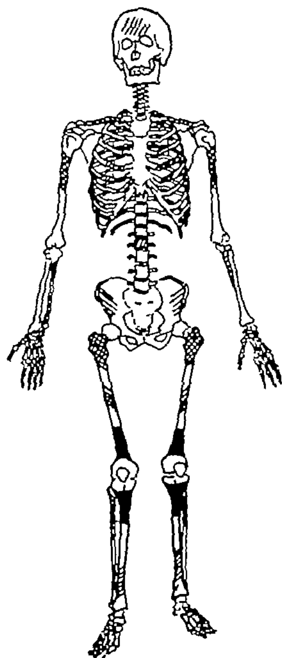
Figure 495 Clinical appearance of patient with osteogenic sarcoma. There is swelling of the metaphyseal area of the distal right femur

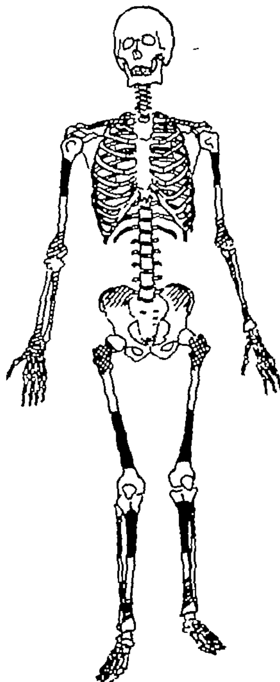


Figure 499 Roentgen features of osteogenic sarcoma metaphyseal location bone forming, globular shape early destruction of the cortex, Codman's triangle of subperiosteal bone at the periphery active destruction and sunburst appearance with radial deposition of bone

is a valid indication to correct treatment may still be argued However patients who have had a delayed amputation with preliminary x ray treatment have had a better than expected course in our direct observation These tumors are felt to be insensitive to x ray therapy There is however a maturing influence since the tumor tends to become more osteoplastic denser by roentgenogram and less painful

The patient gains weight fever if present subsides and elevation of the sedimentation rate if present declines When the tumor is quiescent with no new radiolucent areas the limb is amputated. The level of amputation is not necessarily that of disarticulation but is as far removed from the tumor as possible Recurrence in the stump is rare.





Figures 497, 498. Distribution of sclerosing and osteolytic osteogenic sarcoma with greatest incidence in the metaphyseal areas above and below the knee (shaded black). (*From* Geschickter and Copeland. Tumors of bone. Lippincott, Philadelphia, 1949.)



Figure 499 Roentgen features of osteogenic sarcoma metaphyseal location bone forming, globular shape early destruction of the cortex, Codman's triangle of subperiosteal bone at the periphery active destruction and sunburst appearance with radial deposition of bone

is a valid indication to correct treatment may still be argued However patients who have had a delayed amputation with preliminary x ray treatment have had a better than expected course in our direct observation. These tumors are felt to be insensitive to x ray therapy There is however a maturing influence since the tumor tends to become more osteoplastic denser by roentgenogram and less painful

The patient gains weight fever if present subsides and elevation of the sedimentation rate if present declines When the tumor is quiescent with no new radiolucent areas the limb is amputated The level of amputation is not necessarily that of disarticulation but is as far removed from the tumor as possible. Recurrence in the stump is rare

After amputation the patient is encouraged to follow an active existence rather than bed rest which allows for considerable stasis in the vascular system. Unusual trips or modes of travel are avoided however

References

BADGELEY C. E. AND BATTS M JR. Osteogenic sarcoma. Arch. Surg 43: 541 1941

COLEY B L. Neoplasms of Bone and Related Conditions. Hoeber New York, 1949

FERGUSON A. B Treatment of osteogenic sarcoma. J Bone & Joint Surg 22: 916 1940

LUCK, J V A correlation of roentgenogram and pathological changes in ossifying and chondrifying primary osteogenic neoplasms. Radiology 40: 253 1943

MCNATTIN R. F Treatment of osteogenic sarcoma with preoperative radiation in large doses. Radiology 42: 246 1944

PRATO S R. Clinical analysis of two hundred and five cases of malignant bone tumor J *Bone & Joint Surg.*, 32A: 293 1950

EWING'S SARCOMA

Ewing singled out the sarcoma now called by his name as a clinical entity This was done on the basis of clinical characteristics as well as the pathological findings. He designated it finally as an endothelial myeloma and felt that it arose from the angio-endothelium. It is non-osteogenic and occurs principally in the first two decades of life. It characteristically arises in the diaphysis and spreads axially The metastases are found in other bones and in the lungs. Most series have revealed a preponderance of the cases in males of the Caucasian race, in fact cases in the Negro race are rare It is most common in the tibia or femur

Trauma may precede the development of symptoms by several months, but appears to be no more than coincidental.

Clinical Picture

The usual first symptom is pain localized to the area of involvement. Originally it may be intermittent, but it soon becomes continuous and usually has existed for several months before being seen. This is followed by swelling and the eventual development of a definite mass. The mass tends to be elongated longitudinally rather than localized and in some cases may be the presenting complaint. There is charateristically a febrile elevation usually about 101 accompanying the disease.

If of some duration the sarcoma may simulate osteomyelitis with heat and tenderness of the mass. The vessels are dilated over it, and in some cases fluctuation has been noted The fusiform shape of the mass mimics a late osteomyelitis with subperiosteal diaphyseal abscess.

The mass has been noted to spontaneously regress on occasion only to recur with further symptoms Tenderness of the area is virtually a universal finding.

Laboratory Findings

There is commonly a secondary anemia. Elevation of the leucocyte count is most common. The elevation may be accompanied by an increase in eosinophils, but more commonly the distribution of cells is not unusual. The sedimentation rate is elevated.



Figure 600 Roentgenogram of Ewing's sarcoma exhibiting infiltrative type of destruction tendency to run axially along the shaft multiple layers of subperiosteal ossification A diaphyseal location would be more typical.

Roentgen Findings

The classical case of Ewing's sarcoma is located in the diaphysis It is featured by infiltrative rather than total destruction. Although non-osteogenic, there appears to be an attempt on the part of the host to wall off the invader by bony deposition at the margins of an area of infiltration Individual trabeculae appear to survive as the tumor extends axially along the shaft. There may be mottled areas of destruction as well These areas and the sclerosing reaction may sometimes liken the picture to that of osteomyelitis.

There ordinarily is no difficulty in distinguishing the fact that the lesion is a malignant tumor however although it may be difficult to classify it as a Ewing's Sarcoma

The area of involvement may be overlain with layers of subperiosteal ossification which apparently represent attempts at containing the disease rather than osteogenesis on the part of the tumor itself This "onion skin" appearance though characteristic is not always present The cortical area may be widened and in

areas where the cortex is destroyed there may be minute spicules of bone running at right angles to the shaft. Rarely the tumor arises in the metaphysis, but an epiphyseal origin is unknown. The tumor metastasizes to other bones notably the skull and ilium and to the lungs

Pathology

There appears to be definite resistance to lateral expansion of the tumor which is usually contained by the periosteum. The cortex is thickened by both endosteal and periosteal bone formation. Bone that is infiltrated by tumor appears to be stimulated to lay down new bone to contain the area. The periosteum is elevated and often well separated from the cortex. The vessels are pulled outward at right angles to the shaft by this elevation and aid in determining the laying down of bone in this direction. Successive subperiosteal layering is apparently brought about by repeated hemorrhages

The tumor is encapsulated by fibrous tissue and is lobulated with strands of this tissue extending through it. Areas of hemorrhage may be seen.

Microscopically there are cells with indistinct cytoplasm and round or oval nuclei with mitoses frequently seen. The cells are often quite closely packed

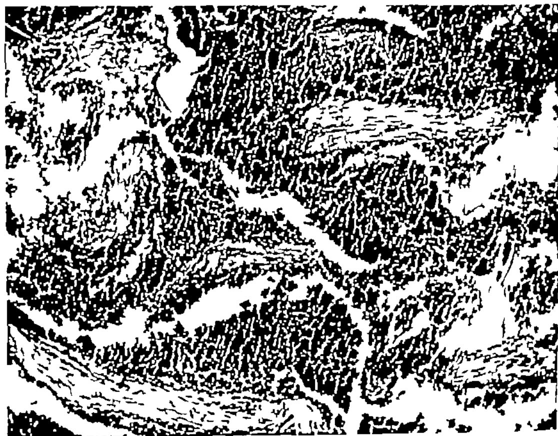


Figure 601 Ewing's sarcoma with sheets of endothelial cells separated by fibrous tissue septa.

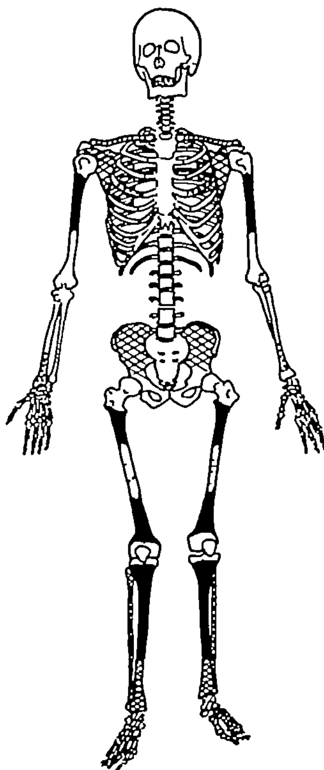


Figure 503 Distribution of Ewing's sarcoma (*From* Geschickter and Copeland Tumors of bone Lippincott Philadelphia 1949)

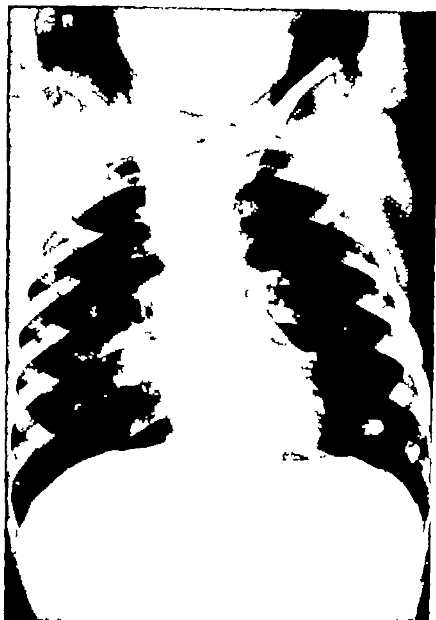


Figure 603 Metastases to the lung from Ewing's sarcoma of the femur

without intercellular stroma These sheets of cells are segregated by occasional fibrous tissue septa.

From pathological examination it is evident that the tumor extends beyond the limits recognizable by roentgenogram. Although the majority of the cases are in the femur and tibia other long bones—the ribs vertebra skull clavicle and small bones of the foot—have been involved

Ewing described this type cell as small and polyhedral with hyperchromatic nucleus and pale cytoplasm and felt that they were derived from angioendothelial tissues



Figure 504 Reticulum cell sarcoma

Prognosis and Treatment

It is evident that the outlook is bleak on reviewing various series. In seventeen cases reported by Lichtenstein and Jaffe which were treated by x ray alone none survived In those reported by Geschickter and Copeland a small percentage survived and radical surgery appeared to give a slightly more favorable outlook.

The principle value of x ray treatment appears to lie in the amelioration of symptoms. The mass characteristically reduces in size following even a short course in treatment

Radical surgery is apparently indicated on the basis of knowledge now available The level of amputation or the possibilities for resection would vary with the anatomic location and clinical features of any individual case.

References

Copeland M M and Geschickter C F Ewing's sarcoma the nature of Ewing's tumor Arch. Surg., **20**: 246 1930

Ewing J Diffuse endothelioma of bone Proc New York Path Soc **21**: 17 1921

Ewing, J A review of the classification of bone tumors. Surg. Gynec Obstet **68**: 971 1939

Lichtenstein L. and Jaffe, H L. Ewing's sarcoma of bone Am J Path., **23**: 43, 1947

RETICULUM CELL SARCOMA

Reticulum cell sarcoma was separated as an entity by Parker and Jackson in 1939 It is believed to be derived from the reticulum cells of the bone marrow and has a more hopeful outlook than Ewing's sarcoma Typically the general condition of the patient is unaffected. It has a tendency to remain localized to a single bone for a considerable time before metastasizing to regional lymph nodes or distantly via the blood stream. The tumor involves adults but some childhood cases have been seen

The roentgenographic picture resembles a Ewing's sarcome somewhat except that it involves the metaphyseal area. It is osteolytic and tends to break out through the cortex on one side. There is little or no cortical or periosteal reaction. It readily involves the epiphyseal region.

This type cell has a round or oval nucleus, approximately twice the size of a lymphocyte. It may be elongated or lobulated There may be a fair amount of cytoplasm. Giant cells are not seen. Mitoses are present. A reticulum stain brings out strands running around areas of tumor cells.

The prognosis is apparently much better than in Ewing's tumor with appropriate treatments This includes amputation or radical resection followed by roentgen therapy

References

PARKER, F JR. AND JACKSON H JR. Primary reticulum cell sarcoma of bone Surg. Gynec. & Obst., 68: 45–53 1939

VALLS, J., MUSCOLO D AND SCHAJOWICZ, F Reticulum cell sarcoma of bone. J Bone & Joint Surg 34B: 588 1952.

NEUROBLASTOMA

A malignant adrenal tumor may present as an apparent primary bone tumor in childhood. A sympathetic neuroblastoma may arise from sympathetic nervous tissue in other areas of the body in addition to the adrenal medulla.

Lesions of the skull are particularly characteristic. Metastases to the long bones are frequently symmetrical bilaterally, but may be isolated lesions of the shaft and confused with a Ewing's sarcoma. Infancy and early childhood is the age group particularly involved, and the young age group is helpful in arousing suspicion of this disease.

The particularly characteristic finding on microscopic study is the arrangement of tumor cells in rosettes Willis has reported on the possibility of confusing a metastasis from a neuroblastoma with Ewing's tumor when the rosettes are not seen

References

TILESTON W AND WOLBACH S. B Primary tumors of the adrenal gland in children. Am. J M Sc 135 871 1908.

WILLIS R. A Metastatic neuroblastoma in bone presenting the Ewing's syndrome with a discussion of Ewing's tumor Am J Path 16: 317 1940

INDEX